THE OPHTHALMOSCOPE

THE ROYAL LONDON OPHTHALMIC HOSPITAL REPORTS
THE OPHTHALMIC REVIEW AND THE OPHTHALMOSCOPE

CONTENTS

Communications:

1. Visual Purple and the Photopic Luminosity Curve. By H. J. Ashton ... 1

2. Scleral Resection in the Treatment of Retinal Detachment. (A Preliminary Report). By Seymour Philips ... 284

3. Fatty Embolism of the Central Artery Found in Eyes after Bandaging and Orbital Exenteration. By Arnold Lorentzen and John Foster ... 289

4. Atropine Cycle Chordotomy at Retinal Surgical Sabs ... 295

5. Intra-ocular Phantoms.—A Report of Three Cases. By Ronald S. Lyster ... 302

Annotations:

1. Results and Causes ... 307

2. Verse or Prose? ... 309

16th International Congress of Ophthalmology ... 311

Book Notices ... 312

Notes ... 317

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THE BRITISH JOURNAL

OF

OPHTHALMOLOGY

NOVEMBER, 1948

COMMUNICATIONS

VISUAL PURPLE AND THE PHOTOPIC LUMINOSITY CURVE*

BY

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Introduction

THE precise relationship which has been established between the scotopic luminosity curve and visual purple (Dartnall & Goodeve, 1937; Wald, 1938) leads one to expect that the photopic luminosity curve is similarly related to some " photopic " pigment or pigments. This latter hypothesis has given rise to considerable speculation, particularly since the evidence for the existence of retinal pigments, other than the various forms of visual purple, is not unequivocal. In any case there is no evidence for such additional pigments in the human retina.

Visual purple mediates scotopic vision by virtue of the photochemical changes it undergoes on exposure to light. The rate of

* Received for Publication, July 12, 1948.

any photochemical change is governed by the value of the product γI_a where γ is the quantum efficiency of the process and I_a the intensity of the absorbed light expressed in quanta per second. Since the eye in the photopic condition is much less sensitive than in the scotopic state it follows that the value of γI_a must be correspondingly smaller for the photopic process.

This low value of γI_a for the photopic process could arise in the following ways:—

(i) γ very small. In this case it is conceivable that the active pigment could be overlooked as being " photostable ".

(ii) I_a very small. This could be due to: (*a*) a very low value for the extinction coefficient or concentration of the active pigment. In these cases detection and isolation would be difficult; or (*b*) the development in a light adapted retina of a stable pigment in a sufficiently high optical density to prevent all but a very slight absorption of light by the active pigment.

The many possibilities are reduced by Granit's (1941) observation with carp and tench retinae. In these retinae, which contain the porphyrhopsin variety of visual purple (absorption maximum at 540 mμ), the photopic curve has its maximum at 600-610 mμ instead of 556 mμ as in the case of retinae with the rhodopsin variety of visual purple. This observation affords strong evidence that the photo-sensitive material mediating the sensation of photopic luminosity is related to the particular form of visual purple possessed by the species.

The simplest assumption accounting for this observation is that visual purple itself is the mediator of photopic luminosity and that the shift in the luminosity curve from its scotopic to its photopic position is the result of other factors which come into play when the eye is light adapted. Thus the bleaching of visual purple results in the formation of yellow substances. The presence of these in a light adapted retina will have two effects. Firstly, there will result an overall reduction in sensitivity owing to the fact that only a fraction of the total light absorbed by the retina will be absorbed by the visual purple. Secondly, because the (yellow) photodecomposition products of visual purple have a heavier absorption in the blue, there will be a relative depression of sensitivity in the short wave end of the spectrum, resulting in a shift of the maximum of sensitivity towards the longer wave end of the spectrum. This possibility of accounting for the Purkinje shift was cursorily examined by Lythgoe (1938). These effects will be enhanced by the fact that the photopic luminosity curve is measured at the central retina which is permeated with the yellow macular pigment, whereas the scotopic luminosity curve is

measured in a more peripheral region of the retina, free from this pigment.

Thus the present hypothesis that visual purple mediates the luminosity sensations of both scotopic and photopic vision offers a *prima facie* explanation of the reduced sensitivity of photopic vision and also of the Purkinje shift associated with light adaptation. This paper is concerned with the quantitative consideration of this hypothesis.

The effect of the presence of photodecomposition products on the light absorbed by visual purple

When solutions of visual purple are exposed to light the red colour fades to a light or deep yellow, depending on the hydrogen ion concentration. Thus in alkaline solutions the colour of the bleached solution is pale yellow, while in acid solutions it is deep chrome yellow. Lythgoe (1937) has called the product of the bleaching with this dependence of colour on pH, " indicator yellow."

At low temperatures (5° C. or less) and in neutral or slightly alkaline solutions an intermediate substance which Lythgoe (1937) has named "transient orange " appears on bleaching visual purple. This substance is rapidly converted to indicator yellow on allowing the temperature to rise.

The retina is stated to become acid on exposure to light and transient orange has not been demonstrated in the bleaching of an acid solution of visual purple. Further, it seems reasonable to suppose that if transient orange were formed under retinal conditions, it would decompose extremely rapidly at blood temperature (37° C.) to indicator yellow. In other words it may be assumed that there is no appreciable concentration of transient orange in a light adapted retina at equilibrium.

The presence of indicator yellow in the light adapted retina will affect the amount of light absorbed by the visual purple and hence also the luminosity curve. The degree of this effect will clearly depend on the amounts of visual purple and indicator yellow present and also on the pH of the light adapted retina since this latter determines the absorption spectrum of the indicator yellow.

The amount of indicator yellow in the light adapted retina. During the process of light adaptation indicator yellow will tend to accumulate in the retina. Lythgoe's work has made it clear that indicator yellow is stoichiometrically related to visual purple, that is, any given amount of indicator yellow corresponds to a definite amount of photochemically changed visual purple. It is also clear from the close relation between the scotopic luminosity curve and

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the absorption spectrum of visual purple that the *dark adapted* retina must be substantially free from indicator yellow and, indeed, free from any substances with non-uniform absorption in the visible spectrum. In consequence, it follows that all indicator yellow present in a light adapted retina is derived from visual purple.

It cannot be assumed from this, however, that the maximum possible amount of indicator yellow in a light adapted retina is merely that resulting from the complete conversion of the " dark " concentration of visual purple as this would imply that the system

$$\text{Visual purple} \underset{\text{light}}{\rightleftharpoons} \text{Indicator yellow}$$

is a " closed " system, *i.e.*, that there are no precursors to visual purple. It will be sufficiently general for the present purpose to assume a scheme such as the following,

$$\text{Precursor(s) of Visual purple} \rightleftharpoons \text{Visual purple} \rightleftharpoons \text{Indicator yellow} \longrightarrow \text{Thermal decomposition}$$

in which the formation of visual purple from precursors is shown as a reversible reaction to account for the fact that visual purple does not accumulate indefinitely in a retina removed from light.

It will be supposed that the production of visual purple from its precursors is a rapid process in comparison with regeneration from its decomposition products. With this assumption, it follows that large quantities of indicator yellow can accumulate in a retina which has been exposed to high intensities for considerable periods provided that the dissipation of indicator yellow by thermal decomposition is not great. This latter condition is satisfied provided the pH is *outside* the range 3·5 to 6·5 (Lythgoe and Quilliam, 1938).

The amount of visual purple in the light adapted retina. After exposure to intense lights for prolonged periods the retinae of frogs may still contain considerable amounts of visual purple (Lythgoe, 1940). This is probably due to the protective action afforded by the migration of the pigment epithelium which occurs on light adaptation. Though this migration is marked in fishes and birds it is not so distinct in reptiles, whilst in mammals it has not certainly been demonstrated. Oguchi (1924) failed to find it in man. Since pigment migration is absent in man it might appear reasonable to suppose that the amount of visual purple is very much less in the light adapted than in the dark adapted state. This, however, does not necessarily follow because indicator yellow also will act as a protective filter, particularly at the shorter wavelengths. If visual purple is rapidly formed from its precursors it

is possible that considerable amounts may be present even in a retina adapted to an intense illumination since, under these conditions the concentration of indicator yellow would be very high and, in consequence, only a small fraction of the total light absorbed would be available for the photochemical decomposition of visual purple. It is also apparent that the concentrations of visual purple and indicator yellow in a light adapted retina will depend not only on the intensity of the adapting light but also on its wavelength distribution of energy, as this latter will affect the degree of internal filter protection accorded by the indicator yellow.

There is no trustworthy measurement of the optical density of the visual purple in a dark adapted human retina. Dartnall and Goodeve (1937) assumed that the value would be similar to that for the dark adapted frog retina and, using extraction data then available for this species, calculated the density at 502 mμ (the peak density) to be 0·1. Broda, Goodeve and Lythgoe (1940) subsequently found, when all precautions against loss were taken, that the density in situ in the dark adapted frog retina is 0·54. It is, however, unlikely that the effective peak density in the human retina can be greater than (say) 0·2 as otherwise the correspondence between the percentage absorption curve of visual purple and the scotopic curve would be lost.

On exposure of a dark adapted retina to light the concentration of visual purple will fall. When the retina has again reached equilibrium (become adapted to the light) the visual purple may regain its former concentration. It is even conceivable that it could exceed it as the presence of a large amount of indicator yellow might disturb the equilibrium between visual purple and its precursors. It does not seem likely, however, that the concentration of visual purple in a light adapted retina would ever greatly exceed its "dark" concentration.

Theory. It will be assumed that the effect of indicator yellow in the retina on the light absorbed by visual purple is identical with that obtaining in a *homogeneous* solution or mixture of these two substances.

Assuming Beer's and Lambert's laws to hold, the fraction of the total amount of light absorbed by any single component of a homogeneous solution containing several absorbing substances is given by the optical density of the single component divided by the total optical density of the solution. Thus in a solution containing both visual purple and indicator yellow, the *fraction* of the total absorbed light of wavelength λ which is absorbed by visual purple is given by

$$\frac{D_\lambda}{D_\lambda + D'_\lambda}$$

where D_λ is the density of the visual purple and D'_λ that of the indicator yellow for light of wavelength λ.

The total fraction of incident light absorbed by the solution is

$$\left(1-10^{-(D\lambda+D'\lambda)}\right)$$

By combining these two terms, the fraction of incident light of wavelength λ which is *absorbed by visual purple* in a solution containing both visual purple and indicator yellow is, therefore,

$$\frac{D_\lambda}{D_\lambda+D'_\lambda}\cdot\left(1-10^{-(D\lambda+D'\lambda)}\right) \quad \ldots\ldots\ldots\ldots\ldots\ldots(1)$$

The assumption that visual purple is produced rapidly from its precursors implies, as previously stated, that considerable amounts of indicator yellow can accumulate during the course of light adaptation. Consider a solution in which the amount of visual purple is kept constant but that of the indicator yellow continuously increased. The effect of the accumulation of indicator yellow will be twofold. In the first place there will be a continuous overall decrease in the amount of light absorbed by the visual purple at all wavelengths. This may be called the "depression" effect since any process which is dependent on the amount of light absorbed by visual purple (such as vision) would be correspondingly reduced in sensitivity. Secondly, because the absorption of indicator yellow is not uniform throughout the spectrum, the amount of this decrease will vary with wavelength. In other words, the *form* of the curve obtained by plotting the amount of light absorbed by the visual purple against wavelength will change. Actually, since indicator yellow absorbs most strongly at the short wave end of the spectrum, the result will be to displace this curve towards the longer wavelengths.

As the amount of indicator yellow increases, the value of the term $\left(1-10^{-(D\lambda+D'\lambda)}\right)$, for the total fraction of light absorbed by the solution approaches its maximum value of unity. In addition the density of visual purple, D_λ becomes less significant in comparison with D'_λ, the density of the indicator yellow. Thus as the amount of indicator yellow increases, the expression for the amount of light absorbed by visual purple approaches D_λ/D'_λ.

As D'_λ tends to an infinitely large value, the amount of light absorbed by the visual purple, D_λ/D'_λ, tends to zero. The "depression" effect thus proceeds continuously with increasing indicator yellow concentration to the limit where no light is absorbed by the visual purple. The "displacement" effect can be isolated from the "depression" effect by plotting the curves

for the wavelength distribution of light absorbed by visual purple as percentages of their respective maxima. The limit of the " displacement " effect is thus given by plotting the values of D_λ/D'_λ, as percentages of the maximum value, against wavelength.

Calculations. Owing to the pH dependence of the absorption of indicator yellow, it is first necessary to arrive at a value for the pH of the light adapted retina. No measurements appear to have been made of the variation of the pH of the retina with adaptation. Lythgoe (1938) has assumed the value 6·5 for the " light adapted " retina. Unfortunately, the absorption spectrum of indicator yellow at this pH is not given by Lythgoe in his 1937 paper, the nearest determinations being at pH 7 and pH 6·1. Provisionally the value 6·1 will be assumed.

The densities of visual purple and indicator yellow solutions at pH 6·1 are shown in the first and second columns respectively of Table I. These figures (Lythgoe, 1937) are the densities of an

TABLE I

The densities at various wavelengths of a visual purple solution and the corresponding indicator yellow solution obtained by photochemical bleaching (columns 2 and 3 respectively) and the values of the ratio of visual purple density to indicator yellow density (column 4). pH 6.1. (From Lythgoe (1937)).

Wavelength mμ	Densities at pH 6.1		Ratio
	Visual Purple	Indicator Yellow	
420	0.35	0.61	0.57
430	0.42	0.54	0.78
440	0.49	0.47	1.04
450	0.60	0.39	1.54
460	0.71	0.33	2.15
470	0.82	0.27	3.04
480	0.92	0.23	4.0
490	1.00	0.18	5.6
500	1.03	0.14	7.4
510	1.01	0.12	8.4
520	0.93	0.09	10.3
530	0.80	0.07	11.4
540	0.65	0.05	13
550	0.47	0.04	12
560	0.31	0.02	15
580	0.12	0.01	12
600	0.04	0.01	4

unbleached and bleached solution and include absorption due to impurities. Lythgoe attempted to estimate the amount of absorption by the impurities, but his methods were not sound and the uncorrected figures are more reliable. It is clear from a study of his paper that his solutions were almost photometrically pure, the density of absorbing impurities being very small, particularly at the longer wavelengths.

The expression (1) for the amount of light absorbed by visual purple consists of two factors. The factor within the bracket evaluates the total light absorbed, that is, by the visual purple and indicator yellow combined; the remaining factor, $\frac{D\lambda}{D\lambda+D'\lambda}$, gives the fraction of this total which is absorbed by the visual purple.

The amount of visual purple in a retina at any given stage of light adaptation cannot, at present, be assessed. It may be very small or it may be of the same order as in the dark adapted retina or, conceivably, even somewhat greater. It depends, among other things, on what degree of protection from bleaching is afforded by the accumulated indicator yellow and this, in turn, depends on the wavelength distribution of the adapting light.

All possibilities will, however, be covered by considering two extreme cases. In the first case the effect of increasing amounts of indicator yellow on the light absorbed by visual purple having a density of 0·5 at 502 mμ will be considered. This concentration of visual purple has been intentionally chosen as being, in all probability, much higher than would ever occur in a light adapted retina. In the second case the effect of increasing amounts of indicator yellow on the light absorbed by visual purple of infinitesimally small peak density will be considered. These two cases constitute upper and lower extremes respectively.

First Case. In Fig. 1 are shown two series of curves. The A-series curves, calculated from the expression $\left(1-10^{-(D\lambda+D'\lambda)}\right)$, show the total fraction of light of each wavelength absorbed by solutions containing (in every case) visual purple of peak density 0·5 and indicator yellow of concentration indicated by the numeral for each curve. The numerals actually represent the peak density of the parent visual purple solution, this being a convenient measure of indicator yellow concentration since visual purple and indicator yellow are stoichiometrically related. Conversion to actual indicator yellow densities may be carried out using table I. Thus from table I and simple proportion the indicator yellow arising from the photodecomposition of visual purple of peak density 10 for example has an actual density of 4·56 at 440 mμ 0·88 at 520 mμ, etc.

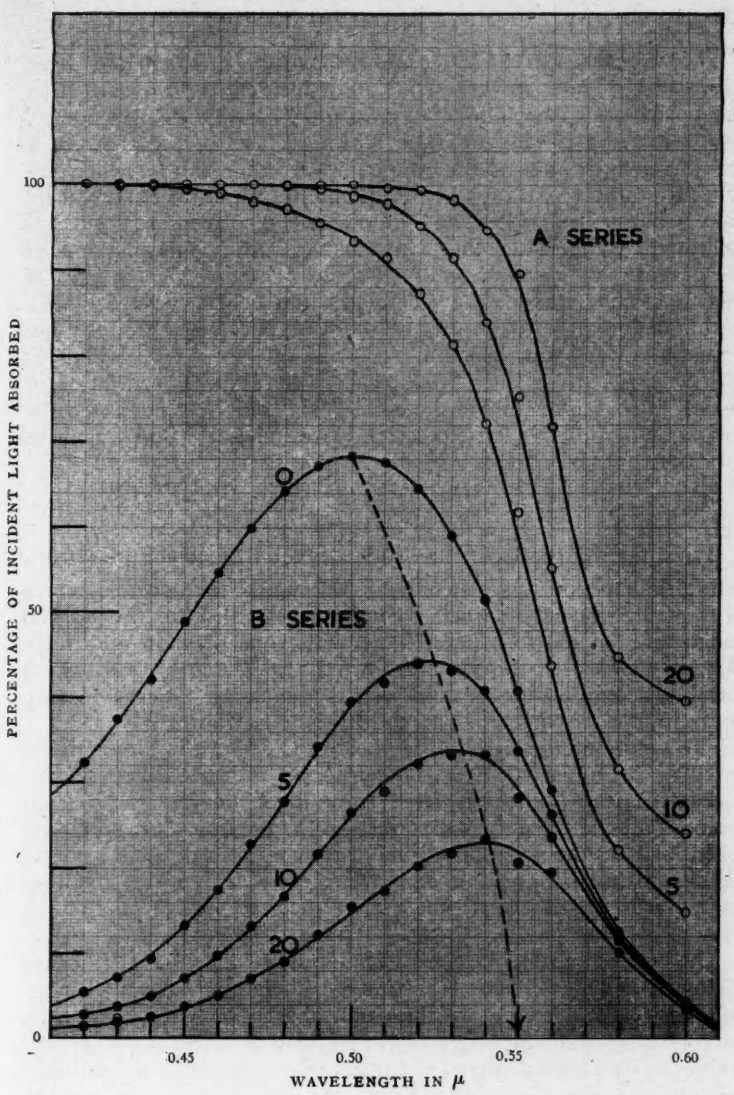

FIG. 1.

A *series curves.*—The total percentage of incident light absorbed by mixtures of visual purple (peak density 0·5 throughout) and indicator yellow of various concentrations. The numerals are measures of the indicator yellow concentrations and refer to the peak density of the visual purple from which they are derived.

B *series curves.*—The "depression" and "displacement" effects, at various indicator yellow concentrations, on the percentage of incident light absorbed by visual purple of peak density 0·5. The curve marked "O" refers to visual purple alone; other numerals are as for the A series curves. The intersection of the dotted curve connecting the maxima of the curves with the wavelength axis indicates the limiting displacement of the maximum at infinite indicator yellow concentration.

The B-series curves of Fig. 1 have been calculated from expression (1) and show the fraction of incident light absorbed by the visual purple. The " depression " and " displacement " effects are clearly shown. The limiting curve of the B-series is the wavelength axis of the graph or, in other words, when the concentration of indicator yellow is infinite the light absorbed by the visual purple is nil. The limit of the " displacement " effect is indicated by the dotted line which passes through the maxima of the curves. By re-plotting the B-series curves as percentages of their respective maxima the " displacement " effect can be isolated. If this is done it is found that the limiting curve of the family has a maximum at about 550 mμ,

Second Case. In this extreme case, where the concentration of visual purple is vanishingly small, D_λ may be neglected by comparison with D'_λ in expression (1) which accordingly may be rewritten

$$\frac{D_\lambda}{D'_\lambda} \cdot \left(1-10^{-D'\lambda}\right)$$

By evaluating the factor $\left(1-10^{-D'\lambda}\right)$ for various concentrations of indicator yellow, a series of curves similar to the A-series curves of Fig. 1 may be obtained. Such curves illustrate the normal broadening of an absorption band with concentration and show that, in the case of indicator yellow, the wavelength up to which virtually complete absorption occurs advances with increasing concentration towards the red end of the spectrum.

The value of the remaining factor, D_λ/D'_λ, is vanishingly small in the present case. Therefore, in order to illustrate the " displacement " effect isolated from the " depression " effect, the ordinates of the curves described in the previous paragraph are multiplied by the appropriate values of the ratio of visual purple density to indicator yellow density from Table I. The resulting curves are plotted as percentages of their respective maxima in Fig. 2.

The values of the ratio of visual purple density to indicator yellow density in Table I are rather uncertain at wavelengths beyond 520 mμ owing to the small absorption of the indicator yellow solution at the longer wavelengths. The resulting irregularities in the derived curves of Figs. 1 (B-series) and 2 accordingly become increasingly apparent as the maximum moves towards the longer wavelengths. Now the *limiting* curve of Fig. 2 is given by the ratio D_λ/D'_λ alone since here the remaining term $\left(1-10^{-D'\lambda}\right)$ is equal to unity. To show the limiting curve of this series more clearly, therefore, it has been plotted from the values for the ratios D_λ/D'_λ of Table I after they have been

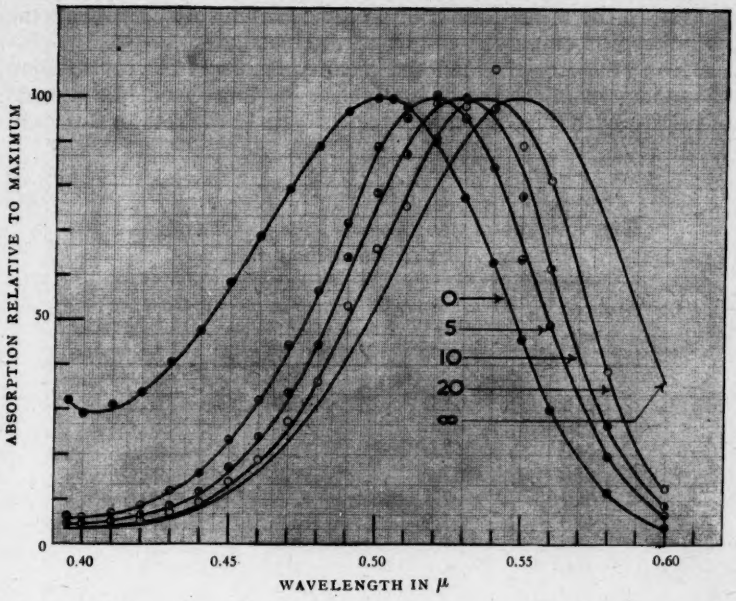

FIG. 2.

The "displacement" effect of various concentrations of indicator yellow on the light absorbed by visual purple of very low concentration. The curve marked "0" is the absorption spectrum of visual purple. The curve marked "∞" is the limiting curve of the series, attained at infinite indicator yellow concentration.

smoothed by the "method of fitting parabolas of degree 2 or 3" (Whittaker and Robinson, 1924).

The results of the calculated examples may be summarized as follows. The absolute value of the "depression" effect (reduction in sensitivity) for a given state of light adaptation cannot be assessed without a knowledge of the concentrations of visual purple and indicator yellow in the light adapted retina. The "displacement" effect (Purkinje shift), on the other hand, proceeds to a limit which is independent of the visual purple concentration. Further, although this limit is attained only when the concentration of indicator yellow is infinite, it is, as shown by Figs. 1 and 2, closely approached at finite concentrations of indicator yellow.

The effect of absorption by the macula lutea and the pre-retinal ocular media

The determination of photopic luminosities is carried out in the central retina where absorption due to the macular pigment takes

place. In the measurement of scotopic luminosities, however, the retinal region employed is beyond the macula.

Sachs (1891) determined the mean coefficients of transmission of monochromatic light by the yellow pigment from the macula of human eyes (nine specimens) and obtained the results shown in Table II.

TABLE II

The variation of transmission with wavelength at the macular region of the retina.

Wavelength mμ	Transmission of macula lutea (per cent.)	Transmission of ocular media (per cent.)	Combined Transmission (per cent.)
420	67.0	16.0	10.7
430	67.0	24.8	16.6
440	67.0	31.8	21.3
450	67.0	38.8	26.0
460	67.1	42.6	28.6
470	67.5	43.8	29.6
480	68.0	45.8	31.1
490	70.0	48.1	33.7
500	74.0	49.5	36.6
510	80.0	51.0	40.8
520	90.5	52.5	47.5
530	94.1	54.0	50.8
540	95.1	55.9	53.2
550	96.2	57.0	54.8
560	97.1	57.2	55.5
580	99.1	59.4	58.9
600	100.0	61.0	61.0

A further source of preferential light absorption is that by the aqueous, lens, and vitreous. This absorption is, of course, always present irrespective of the retinal region investigated. Ludvigh and McCarthy's data (1938) for the transmission of the ocular media are also shown in Table II.

In the last column of the table the combined effect of these two factors is shown.

Derivation of the photopic luminosity curve

In order to complete the comparison with the photopic luminosity curve, the ordinates of the curves of Figs. 1 (B series) and 2 require to be multiplied by the transmission factor appropriate to each wavelength (shown in Table II). The families of curves so

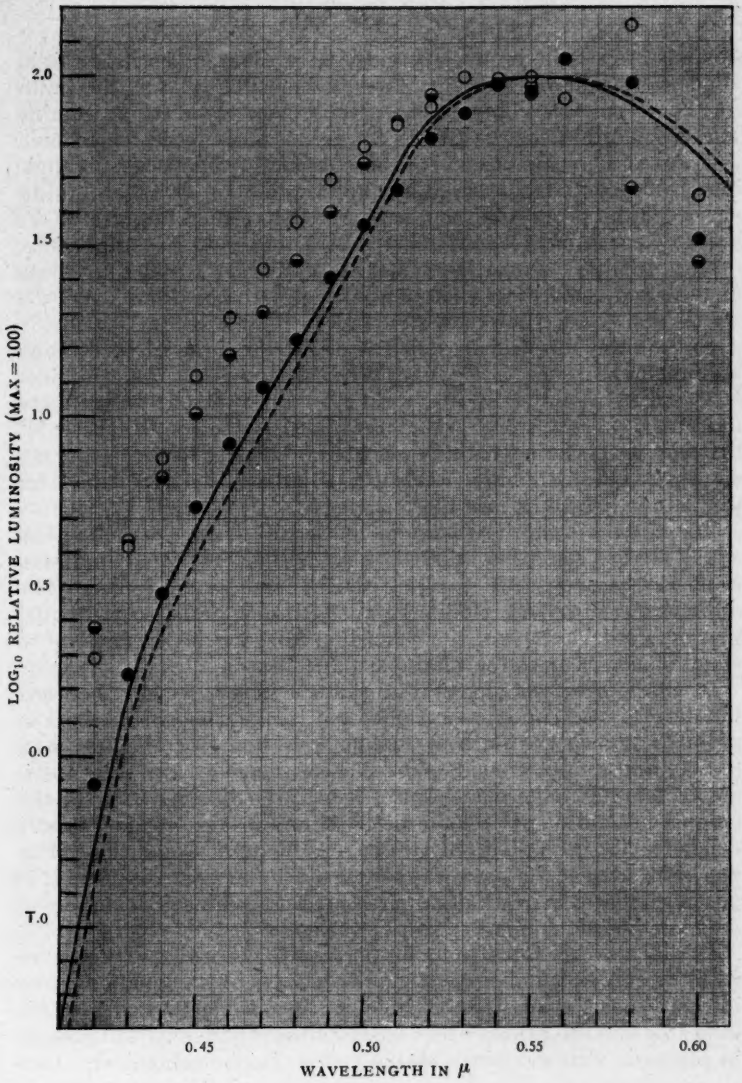

FIG. 3.

Experimental and calculated photopic luminosity curves.

– – – – luminosity curve (experimental) on the basis of an equal energy spectrum.

——— ditto on the basis of an equal quantum intensity spectrum.

○ calculated values assuming pH = 7.0.

● ditto assuming pH = 6.1.

◒ ditto assuming pH = 5.2.

obtained represent luminosity curves for different levels of light adaptation (different concentrations of indicator yellow), the limiting curve corresponding to the condition of complete adaptation to a very high intensity (infinite concentration of indicator yellow). As this latter condition is approximated when the photopic luminosity curve is measured it is appropriate to select the limiting curve for the purpose of comparison. When the ordinates of the limiting curve (the ratios D_λ/D'_λ of Table I) are multiplied by the appropriate transmission factors of Table II and the values obtained adjusted proportionately so that the maximum is 100, the values shown by filled circles in Fig. 3 are obtained.

In the foregoing calculations the pH of the light adapted retina was assumed to be 6·1. This is probably rather on the acid side. If the pH is taken as 7·0 and the value of D_λ/D'_λ for this pH are multiplied by the appropriate transmission factors of Table II, the luminosity values for this pH are obtained. These results are shown as unfilled circles in Fig. 3. Similarly derived values for pH 5·2 are shown as half-filled circles.

For comparison, the photopic luminosity curves based on an equal energy spectrum (dotted line) and on an equal quantum intensity spectrum (full line) are shown in the same figure. The ordinates of Fig. 3 are plotted on a logarithmic basis, so that equal percentage deviations of the calculated luminosities correspond to equal distances from the photopic curves.

It will be observed from Fig. 3 that the best agreement between the derived and experimental photopic luminosities is obtained at pH 6·1. Since the derived luminosities both for the less acid (pH 7) and the more acid (pH 5·2) cases deviate from the experimental curve in the same sense, it may be inferred that in the neighbourhood of pH 6·1 the derived luminosities are not closely dependent on pH. Thus Lythgoe's estimate of 6·5 as the pH of the light adapted retina is supported by the present results. It is possible that the variation between the luminosity curves of different individuals is due to pH variations.

It will also be noted that the derived luminosities agree better with the experimental curve when the latter is on an equal quantum intensity basis than when it is on an equal energy basis. This indicates that the primary part of the physico-physiological process in photopic vision (as in scotopic vision) is photochemical.

Discussion

The foregoing derivation of the photopic luminosity curve rests on two main assumptions. These are:

1, that the production of visual purple from precursors is a rapid process in comparison with regeneration from its photoproducts;

2, that the influence of indicator yellow on the light absorbed by visual purple is that which would obtain in a *homogeneous* mixture of the two substances.

The first assumption allows the possibility of the accumulation of relatively large amounts of indicator yellow in a light adapted eye, provided that thermal dissipation is not too rapid. (At pH 6·5 this condition is satisfied.)

Insufficient is known about the production of visual purple from precursors to discuss this assumption profitably. It is, however, possible to make a rough estimate of the minimum amount of indicator yellow required to shift the *maximum* of the derived luminosity curve to its photopic position (550 mμ). The validity of the ratio D_λ/D'_λ as a measure of the fraction of incident light absorbed by visual purple, depends on the assumption that *all* the incident light is absorbed. Strictly speaking, this is true only when the density of the indicator yellow is infinite. Absorption is, however, complete for practical purposes (>95 per cent.) at densities of 1·3 and more. As the concentration of indicator yellow increases in the retina, the point in the spectrum up to which there is virtually complete absorption moves towards the longer wavelengths. The maximum of the limiting curve of Fig. 2 is at 550 mμ (approx.). Consequently when the concentration of indicator yellow becomes such that its density at this wavelength is equal to or greater than 1·3, then the derived luminosity curve will also have its maximum at this wavelength and will, moreover, correspond exactly with the limiting curve on the blue side of the maximum. Further increase in the concentration of indicator yellow will result in a raising of the ordinates on the red side of the maximum to their ultimate positions corresponding to the limiting curve of Fig. 2.

Now the photodecomposition of visual purple of peak density 1·0 yields indicator yellow of density 0·04 at 550 mμ at pH 6·1 and 0·05 at pH 7. Taking the mean density, 0·045, as applicable to a pH of 6·5 it is seen that to achieve a density of 1·3 would require the decomposition of visual purple of peak density 1·3/0·045= ca. 29. [This calculation refers to the extreme case where the concentration of visual purple in the light adapted retina is regarded as very small. If the other extreme (peak density 0·5) is assumed then the corresponding visual purple density at 550 mμ, namely, 0·23 must be subtracted from 1·3 since it contributes to the total density. In this case the amount of indicator yellow required is that derived from visual purple of peak density 1·07/0·045=ca. 24.]

These can only be regarded as rough estimates since the absorption of indicator yellow at 550 mμ is not very accurately known. They

indicate, nevertheless, that a considerable amount of photodecomposition of visual purple is necessary to produce sufficient indicator yellow to account for the whole of the Purkinje shift. It is for this reason that the first assumption has been made, namely that the production of visual purple from precursors is a rapid process. This is in distinction to the generally held view which regards the precursors as merely making good the inevitable katabolic losses of visual purple and its photoproducts.

Information concerning the mode of occurrence of visual purple in the retina is insufficient to allow a discussion of the likelihood of the second assumption, namely that the division of absorbed light between visual purple and indicator yellow is such as would obtain in a *homogeneous* mixture of the two substances. In support of this assumption, however, is the fact that the displacement of the luminosity curve as light adaptation proceeds, that is, as the amount of indicator yellow accumulates, is not an indefinite process. *The limit beyond which no further change takes place corresponds to the photopic luminosity curve.* If, as an alternative hypothesis, it were assumed that the indicator yellow acts as an external light filter, its effect, as it accumulated in the retina, on the light absorbed by visual purple and hence on the calculated luminosity curve would be to shift the maximum further and further into the red.

If the above two assumptions are allowed then a number of visual phenomena are capable of explanation. Thus, the failure to find a photosensitive pigment corresponding to the photopic luminosity curve is due to the dual role of visual purple in acting as the mediator of both scotopic and (in the presence of indicator yellow) photopic luminosity. The Purkinje shift is *quantitatively* accounted for, the photopic luminosity curve being calculated as the *limiting* case of the effect of indicator yellow on the light absorbed by visual purple. The corresponding shift in the case of retinae containing porphyrhopsin is similarly explained. Further, the derived photopic luminosity curve agrees better with the measured curve when the latter is plotted, in accordance with photochemical principles, on an equal quantum intensity basis. The lower sensitivity of the light adapted retina is at least partly accounted for by the screening action of the accumulated indicator yellow.

A basis for the explanation of certain aspects of dark adaptation is also provided. As is well known the course of dark adaptation, as measured by the visual threshold at various intervals of time, depends on the previous history of light adaptation. For example, after exposures to light for short periods the rate of return of sensitivity is much more rapid than after exposures for long

periods even though the thresholds immediately after adaptation may be similar.

The presence of variable amounts of indicator yellow in the light adapted retina is consistent with these facts. Thus while a short exposure may be sufficient to reduce the visual purple concentration (and hence the sensitivity) to a low figure, the total amount of photodecomposition during the adapting period and hence the amount of indicator yellow formed may be relatively small. In consequence, on return to the dark, the concentration of visual purple will be rapidly restored from precursors and indicator yellow, any residual amount of indicator yellow being small and hence without great effect on the sensitivity. After exposures for long periods, however, the total amount of photodecomposition, and hence amount of indicator yellow formed, will be large. Consequently, although on return to the dark the concentration of visual purple will be equally, if not more, rapidly restored, indicator yellow will still at this point be present in large amount. Not until this " excess " has been discharged will the sensitivity of the retina return to its full value corresponding to complete dark adaptation.

The phenomenon of " rod monochromatism " (Wright, 1946) can also be accounted for. Subjects with this visual defect are unable to discriminate hues and, while having a normal scotopic luminosity curve, suffer from nystagmus, photophobia and poor visual acuity under conditions of illumination which, in a normal observer, would evoke photopic vision. An additional characteristic is an abnormally slow rate of recovery (dark adaptation) following exposure to a bright light. As the name implies, the usual explanation given for this type of defect is that vision is mediated by rods alone, cone vision being absent. This is little more than a description however. On the present hypothesis the absence of a photopic luminosity curve and the slow rate of dark adaptation are attributable to a reduction in the capacity for the production of visual purple from precursors. Thus the absence of a steady supply of visual purple would prevent the accumulation of sufficient indicator yellow in bright light to displace the luminosity curve to its photopic position. The photophobia presumably arises from the absence of the normal protective action afforded by the indicator yellow.

In the light adapted condition the eye is colour sensitive. It is outside the scope of this paper to discuss the mechanism of colour perception; it is mentioned merely for the purpose of clarifying the implications of the present concepts. The sensation of luminosity is generally regarded as an intensive property of the sensation of hue. While there are practical reasons for this there

are many phenomena, a number of which are given by Troland (1922), which indicate that the sensations of luminosity and hue are mediated by separate processes. The existence of a defect known as " cone monochromatism " (Pitt, 1944), in which the subject has normal dark adaptation, normal scotopic and photopic luminosity curves and yet no colour sense, is sufficient indication that luminosity and hue sensations are not necessarily related as intensive and extensive attributes.

One of the many difficulties of the Duplicity Theory is in accounting for the rôle of the rods at high illuminations. This difficulty arises directly from the habit of regarding luminosity as the intensive property of hue. If the present hypothesis that visual purple mediates both scotopic and photopic luminosities is true, then it is possible that the rods are responsible for the mediation of luminosity both at scotopic and photopic levels, leaving the cones for the mediation of the hue sensations.

Summary

The hypothesis that visual purple is the mediator of photopic luminosity sensations as well as of scotopic is investigated. Two main assumptions are made in developing the hypothesis. The first of these is that the production of visual purple from its precursors is a rapid process in comparison with regeneration from its photoproducts; the second, that the influence of absorption by the photoproducts on the light absorbed by visual purple is equal to that obtaining in a homogenous mixture of the substances.

It is shown that the effect of the accumulation of photoproducts (indicator yellow) in a retina exposed to light, is to move the position of maximum light absorption by visual purple towards the longer wavelengths. This process is not indefinitely prolonged. As the amount of indicator yellow accumulates the light absorbed curve of visual purple rapidly approximates to a *limiting* position having a maximum at about 550 mμ. When this limiting curve is corrected for absorption by the ocular media and by the macular pigment, the resulting curve closely approximates to the photopic luminosity curve.

Apart from accounting for the Purkinje shift in a quantitative manner the hypothesis provides a basis for the explanation of a number of other visual phenomena, notably the reduced sensitivity of photopic vision and the dependence of the rate of dark adaptation upon the previous light history of the retina.

Acknowledgment. I wish to record my thanks to Miss M. A. Hooper for assistance with the computational work involved in the preparation of this paper.

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SCLERAL RESECTION IN THE TREATMENT OF RETINAL DETACHMENT

(A Preliminary Report)

BY

SEYMOUR PHILPS

LONDON

THE great majority of retinal detachments are cured by Gonin's operation of scleral diathermy over the site of the retinal tear, but there remains a percentage which is not cured by this means. This percentage varies from 20 to 30, and the causes of failure are various. Apart from those patients in whom inter-current disease or other factors make operation impossible, the causes of failure of the diathermy may be:—

(1) Failure to see and therefore to seal off the retinal tear.

(2) Retraction of the vitreous and shrinkage of the retina, making it too small to "fit" the eye, so that even after a successful diathermy operation the retina breaks away again.

Failure to see the Retinal Hole. If the media are transparent the whole of the retina can be examined ophthalmoscopically and a hole, if present, can be located. Lack of transparency of the media whether due to lens opacity, capsular remains or vitreous opacity may mean that the view of the retina is so poor that all efforts to locate a retinal tear are fruitless.

A " blind " diathermy operation is rarely successful, and there is a definite limit to the area of scleral surface which can be diathermised without causing undue reaction in the eye, thrombosis of the vortex veins and a choroidal exudate and haemorrhage which makes the last condition of the patient worse than the first.

If one blind operation of limited extent over the supposed site of the hole is unsuccessful, it is better to carry out scleral resection than to repeat the diathermy.

Retraction of the Vitreous and Shrinkage of the Retina. Diathermy operations can only succeed if the retina can be got into contact with the choroid and remain there long enough for scar tissue to form. If, owing to the length of time the detachment has existed, or to other causes, the retina has contracted and shrunk, diathermy will be unsuccessful. Retraction of the vitreous in high myopia or aphakia will have the same effect.

The operation of scleral resection is designed to help those patients in whom the Gonin diathermy operation has failed, and who must otherwise become blind. It is not intended in any way to replace the diathermy operation, though it sometimes offers a better chance of cure where opacification of the media makes retinal examination impossible.

Where the retinal tear can be seen and treated the diathermy operation is the operation of absolute choice and should always be employed first. But where the lens opacities, capsular remains or vitreous opacities obscure the view scleral resection may be employed. Detachment of the retina after extracapsular extraction is an especial indication, as the capsule will almost certainly obscure the view of the peripheral retina and the site of the retinal tear cannot be located.

Indications. (1) Detachments with multiple rents and thin atrophic retina where diathermy has failed, or where the retina breaks away extensively after a successful diathermy operation.

(2) Detachment where no rent can be found because of obstruction to retinal examination.

(3) Long standing detachments where the retina has shrunk and will no longer fit the globe.

(4) Some detachments in very high myopia where the vitreous retraction and thin retina give little chance of success with diathermy.

Rationale of the Operation. Scleral resection shortens the eye and this allows a shrunken retina once more to fit the globe; it also collapses the peripheral retina which contains the rents or degenerative patches which have caused the detachment. It may be employed round the whole 360° of the globe though not more than 180° should be attempted at a single operation, and the

surgeon should aim to place the centre of the scleral arc he resects over the probable site of the retinal hole.

The operation takes time and patience but is not difficult, except perhaps when resecting those areas where repeated diathermy cauterisations have been employed. Certainly the operation is less difficult than those might think who have not employed it.

Anaesthesia.—Local instillation of cocaine 2 per cent. and adrenalin 1: 1000. Retrobulbar injection of novocaine 2 c.c.

Instruments.—Speculum (Lang's). Knife (Bard Parker No. 15 blade, ground down Graefe knife). Scissors (Fine, straight, pointed). Hooks (Double and single conjunctival squint hooks). Needle holders. Forceps (Conjunctival and fixation). Calipers. Measuring Rule. Ink Compass.

Sutures.—1. Traction—Maddox cataract needle 00 black silk. 2. Conjunctival—No. 6 eye curved (two) No. 1 black silk. 3. Muscle—No. 6 eye curved (two) 00 white silk. 4. Scleral—8 double-armed Maddox cataract 00 white silk.

No special instruments are required for the operation, but the needles used must be good ones and should be tested carefully beforehand. A good Maddox cataract needle is required for scleral suture, though perhaps the Grieshaber corneal needle is best of all.

Preparation of Sutures for Scleral Resection.—The suture material used for the sclera should be 00 white silk and the method of preparing silk sutures in general and these in particular needs some explanation. The needles should be gold plated, as this prevents rust and gives a smoother surface which slides better through tough tissues. They are threaded by the theatre sister and each suture is tied to the needle with one half hitch. The sutures are then threaded into the non-fluffy side of a square of white lint, and when sufficient needles have been threaded for the operation the lint is folded into a packet and placed in the dressing drum along with towels, swabs, etc., to be autoclaved.

After sterilisation this suture packet is placed on the surgeon's tray and opened out ready for use. As each suture is required the surgeon picks it out of the lint with the needle holder and inserts it into the sclera.

The advantages of this method are as follows:—

(1) The sutures are handled by no-one. Their sterility is thus assured.

(2) They are dry and easy to use.

(3) They are under the surgeon's eye who picks them up as needed with no words spoken and no turning round to take them from another tray at his side or back; a great advantage when a number of sutures have to be inserted.

TECHNIQUE

Preparation of the Eye. The eyelids and facial nerve should be infiltrated with novocain and adrenalin to give a large palpebral fissure and the maximum space in which to work; after which a traction suture is inserted through episclera close to the limbus and the eye rotated to one side and fixed in that position. By this means it is possible to bring the area of resection into the centre of the operative field.

Exposure of the Operative Field. The conjunctiva is divided throughout the whole length of the planned resection and a rectus muscle detached from its insertion after being picked up with silk sutures. It will now be possible to take the eye over a little further by means of the traction suture and so gain more space for the resection.

Marking out the Resection. The resected sclera should be about 4 mm. wide and the strip should extend round half the circumference of the globe, tapering a little at each end. The site of the resection must be over the peripheral retina between the equator and the ora serrata, and the anterior cut is made 10 mm. from the limbus.

The lines of these incisions are marked out by a series of dots made with a sterile ink drawing compass, the sclera being well dried first to prevent the ink from running. When the incisions have been clearly marked out the resection may begin.

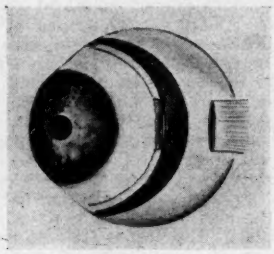

FIG. 1.

The size and shape of the scleral crescent to be resected showing the relation to the rectus muscle insertion.

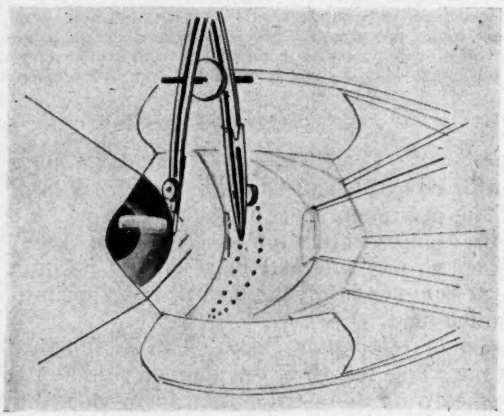

FIG. 2.

Marking out the area to be resected with an ink compass. The two lines of dots are at 10 and 14 mm. from the limbus.

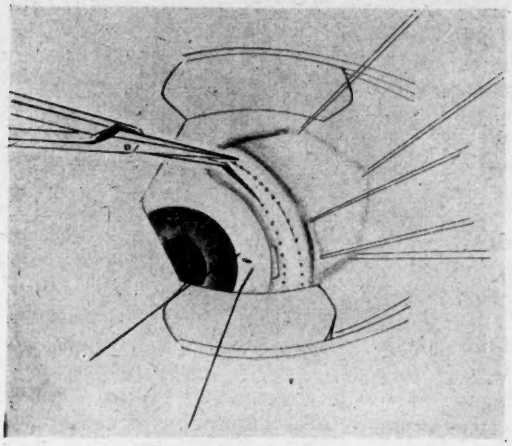

FIG. 3.

Method of starting the resection.—A knife cut has been made through the sclera and the incision is extended from that point along the posterior dotted line with fine straight scissors.

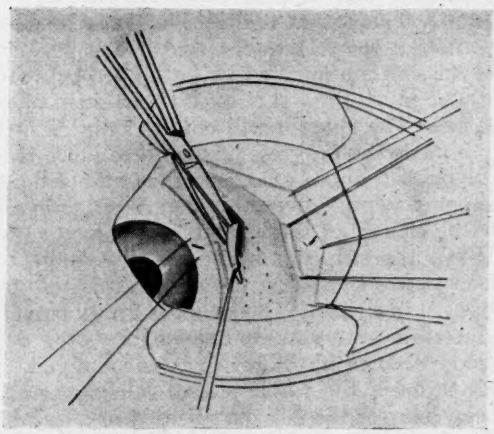

FIG. 4.

Continuation of the resection by means of scissors. The scleral arc is retracted with a hook.

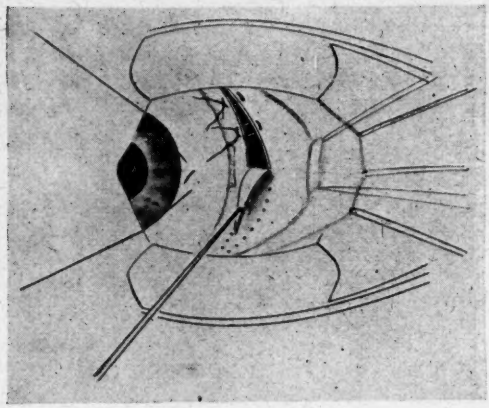

FIG. 5.

Method of inserting the sutures as the resection proceeds.

The Scleral Resection. The surgeon should be seated in such a position that the cutting instrument comes naturally to the right hand. Starting at one end of the scleral crescent the incision is begun with a knife—a Bard Parker is convenient (No. 15 blade)—by making a cut through the sclera until the shining black choroid shows through. The rest of the resection may be continued with good pointed straight eye scissors, and it will be found that one blade of the scissors can quite easily be slipped between sclera and choroid without puncturing the latter and a strip of sclera raised in this fashion. The choroid will tend to bulge a little, but does not usually impede the progress of the operation. If it should do so it may be pricked and a quantity of subretinal fluid evacuated. In any case, this step is advisable at some stage in the operation to facilitate the closure of the wound, and it may as well be early as late. When the strip has been cut for a length of 5 mm. a double-armed suture should be inserted so that too much choroid is not exposed at one time. The sclera is firmly fixed with forceps before the insertion of this suture to avoid any drag on the tissues, but with good needles no difficulty is encountered. The suture should pass through the whole scleral thickness and when each limb has been inserted the silk may be tied in a half hitch and the wound edges drawn together. By continuing this process of cutting and suturing the whole 180°, or whatever resection is planned, may be accomplished without great difficulty, except sometimes in an area where repeated diathermy operations have

been performed. In such places the choroid may be adherent and more difficult to separate from sclera, but it can always be achieved with care and patience. In other parts the scissors slip easily along the suprachoroidal lymph space.

When all the sutures have been inserted the surgeon goes back to the beginning of the resection and, tightening each suture a little, completes the knot and cuts off the ends. With replacement of the rectus muscle and suture of the conjunctiva the operation is completed.

Atropine is instilled and both eyes are bandaged.

Post-Operative Treatment. This is much as for retinal detachment. The patient should be semi-prone on the back in a position of comfort, and the first dressing can be undertaken after two clear days have elapsed from the time of the operation. Binocular bandages are retained for a fortnight and the patient is got up three weeks after the operation.

Post-Operative Course. In a successful case the detachment is much improved when first examined at the end of one week, but there will usually remain some small amount of detachment for some weeks which slowly disappears. The reaction from this operation is much less than might be supposed from its extensive nature, and usually is no more than might be expected after a diathermy operation.

CASE REPORTS

(1) J.T., aged 59 years, London labourer. Presented himself at the Moorfields, Westminster, and Central Eye Hospital having lost the sight of the right eye, which was his only useful eye, following an accident two days previously.

Past History.—Ten years before, he had developed a cataract in the left eye and since that time had depended on the right eye, which was good. Two days before admission, while demolishing a building, a piece of concrete struck him in the right eye, the sight of which deteriorated over the next two days.

Examination.—The patient was a big heavy man, six foot tall and sixteen stone in weight. Right eye, vision 6/36, vitreous haemorrhage obscured many fundus details but there was a detachment of the outer half of the retina, no hole being found owing to the turbid vitreous humour. L.E. Vision = Perception of light. There was a mature cataract, but light projection was accurate and quick.

Treatment.—The patient was put to bed in binocular bandages but in spite of this the right retinal detachment increased and operation had to be undertaken under most unfavourable circumstances over the probable site of the retinal hole.

After two diathermy operations the right retina was even more detached than before and lens and vitreous opacities prevented any adequate fundus view. It was therefore decided to remove the left cataract, which was accordingly done by the intracapsular method on April 12, 1947. In spite of a very good operative result, the retina was found to be detached centrally, below, and to the outer side—very atrophic and the best vision less than 6/60. By this time the right retinal detachment was total and the media opaque, and the only hope of preserving any useful vision lay in improving the left eye. In this eye the retina on the temporal side was all so thin and lacelike that it was difficult to say which areas contained holes and which were merely atrophic. A diathermy operation all down the temporal half of the globe was unsuccessful, and the man finally left hospital with so little vision that he could not see his way about; but on reading a report in

the American ophthalmic literature of the scleral resection operation[1], it was decided to re-admit him and try to improve the left eye by this means. At this time 3/4 of the left retina was detached, the only portion remaining attached was between 10 and 12 o'clock. It was still thought most probable that the original tears causing this detachment were situated above and temporally and so a scleral resection of the outer half of the globe from XII to VI was planned as a first-stage. This was performed on January 10, 1948, after the method described, and the patient was discharged on February 15, 1948, with the retina in place.

The patient was last seen in July, 1948, with the retina still in place. Although, owing to the size of the detachment which had been present, the vision was not more than 5/60 the field was quite good and the patient could see his way around by himself. The line of the scleral resection can be seen quite clearly all down the outer half of the fundus on ophthalmoscopy.

(2) E.B., aged 10 years, London schoolgirl. Was brought by her mother to St. Bartholomew's Hospital on March 20, 1947. The child's left eye had been found to be defective and in spite of the fact that the mother had been advised that this eye was lazy and had been defective since birth, she (the mother) decided to seek other advice as, having herself suffered from retinal detachment, she thought it possible her daughter might suffer from it also.

Past History.—The girl had had a blow in the left eye with a stone three years previously. She had no idea when the left vision began to fail as the defect was discovered during a routine school examination.

Examination.—Right Eye 6/6 unaided—Normal. Left Eye less than 6/60—Counts fingers at one foot.

The left retina was extensively detached, the whole temporal half of the retina including the macula being separated. A disinsertion extended from 11.30 o'clock right round the temporal periphery to 5.30, the area of greatest disinsertion being in the 2.30 meridian.

On March 25, 1947, a diathermy operation was performed and was successful in replacing the retina. The girl left hospital on April 15 with a full field and vision of 4/60. The retina stayed in place until January, 1948, when it suddenly broke away from the scar and once more a long ragged retinal tear appeared from 2 to 5 o'clock, the whole retina being now detached. It was felt that a further diathermy was unlikely to be successful as the retina had retracted so far away from the choroid, and in order to prevent this eye becoming quite blind and undergoing the secondary changes and complications usual in an uncured detachment, a scleral resection was undertaken in February, 1948, to bring the retina nearer to its bed and enable it to become attached again. This resection was from 12 to 5.30 o'clock and the area resected was that strip between 12 and 15-16 millimetres from the limbus. Following this the retina settled in place and has remained so since that time. The vision is poor as the macula was detached, but the media are clear, the eye quiet and cosmetically normal.

This preliminary report is intended to show that the operation has possibilities and is worth serious consideration in those patients where diathermy has failed and who will otherwise go blind.

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FATTY EMBOLISM OF THE RETINAL ARTERY FOUND IN EYES AFTER ENUCLEATION AND ORBITAL EXENTERATION*

BY

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WHILE studying the retina with the slit-lamp after equatorial opening of the excised eye, we have observed, on four occasions, the central artery to be partly filled with fat.

The first case was a malignant melanoma of the choroid in the macular area; the second a malignant melanoma of the limbus; the third a perforating injury of the eyeball; and the fourth an exenteration of the orbit for malignant melanoma.

In three cases the eye was enucleated, and in the fourth exenterated with the orbital fat. All specimens were fixed in 10 per cent. formalin immediately after removal.

The artery appears as a shining white band (Fig. 1) rising from the depth of the cup, while the veins may be empty or filled with

FIG. 1.

Malignant melanoma of the limbus. The central artery and the main branches are filled with a smooth whitish mass. ×5 approx.

FIG. 2.

Malignant choroidal melanoma in macular area. Lower papillary artery filled by a smooth whitish mass. ×2½ approx.

* Received for publication, June 4, 1948.

blood. The higher magnification of the corneal microscope shows interruptions of the milky white content of the arterial branches, whose peripheral twigs are either bloodfilled or collapsed. The white band was visible in another case (Fig. 2), $1\frac{1}{2}$ d.d. only from the disc, while the remaining vessels did not reveal anything abnormal, either in their content or in the walls.

The third case, the perforating injury, showed a short white band only, while the arterial system of the fourth case was milky

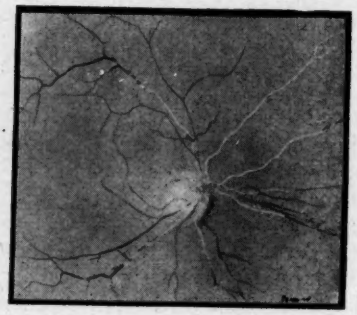

FIG. 3.

Case 4. Malignant melanoma. Exenteration of the orbit.

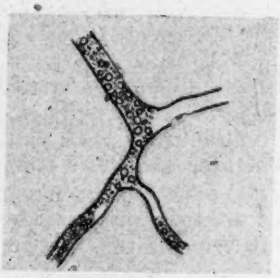

FIG. 4.

Embolism, unstained retina in bulk. oil immersion. A capillary filled with an emulsion of droplets. They are of various sizes (from 2μ down).

to a considerable extent (Fig. 3). The area of transition from the milky part to the bloodfilled lumen is empty for a short distance.

In some places the interrupted peripheral blood column resembled the " cattle truck " phenomenon seen ophthalmoscopically at the instant of death. Here we ought to mention that " cattle trucking " in the retinal blood vessels cannot be regarded as absolute proof of the death of a person (as a writer to the *Brit. Med. Jl.* has assumed lately erroneously), but is also observed in thrombosis of the central retinal vein, or of its branches in the living eye.

The first task was to establish the nature of the vascular content. A disc of the retina with a piece of the milky vessel was excised with a 4 mm. trephine, cleared with glycerine, and studied with high power (Fig. 4). The white vessel was filled with a fine granular mass. The size of the granules varied from 2μ downwards, the great mass of them being far smaller. The majority were doubly refractile. No red blood corpuscles were found within the granular content.

The vessel distal to the " milky " appearance contained blood.

The endothelial cells appeared normal under oil immersion. None of the four patients suffered from diabetes or lipaemia. In no case was an embolus of the central retinal artery seen prior to excision.

The bulk specimen was washed and stained for fat with scarlet red. The whole milky content was shining red, the vessel wall appeared fat free in the fat stained bulk specimen.

Another piece of retina 4 mm. dm. was embedded in gelatine and cut with the freezing microtome and the sections stained with hematoxylin and sudan III. The fatty content appeared darker red (Fig. 5). In spite of careful handling the content of the artery fell out of several cross sections of the optic nerve in the specimen.

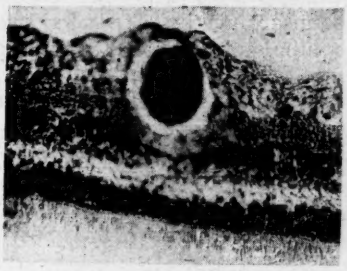

FIG. 5.

Case 4. Retinal disc 4 mm. dm. removed, studied in bulk unstained. Embedded in gelatine, sectioned with freezing microtome stained with haemat., Sudan III. ×300. The retinal artery is filled with a dark-reddish granular mass.

We came to the conclusion that all four cases could be explained by entry of a fatty embolism into the central artery. As the first cases were performed while we were using an anti-rusting substance, A.C. 10, in sterilisation, our attention was first drawn to this. This conception had to be abandoned, as A.C. 10 was not employed in the third case.

We concluded, therefore, that it was orbital fat which had intruded into the divided central artery. The scissors which had cut (unintentionally) orbital fat, had opened the central artery, probably where it pierces the optic nerve sheath (7-12 mm. behind the bulb). The fat was moved by the pulse wave of the artery towards the branches, and was emulsified by the arterial blood. The time lag between intrusion of the fat into the retinal artery, and excision and fixation of the bulb, was too short to permit of pathological changes in vessel walls and retinal tissue.

Fat embolism in the retinal vessels is not as rare as Leber (1915) in his classical description has assumed. According to him no case

has been published in which retinal function has been damaged by fatty embolism.

Urbanek (1933), Oppolzer (1934), Loewenstein (1936) have proved that especially after single or multiple fractures, involving the medulla of the long bones, not only is pulmonary and cerebral fatty embolism relatively frequent, but that retinal changes of typical appearance can be observed.

Urbanek has demonstrated that haemorrhages are as a rule the first sign, and are followed by white patches. The white foci are predominantly situated at the bifurcations of arteries, and they change their position. Histological investigation proved that retinal and choroidal capillaries are partly filled with fat. Urbanek assumes that fatty droplets pass from the bloodstream into retinal tissue, and can disappear quickly. He called attention to the great similarity of the ophthalmoscopic picture of Purtscher's traumatic retinal angiopathy to fat embolism of the retina.

Loewenstein (1936) collected four further cases in a short time. He stressed the fact that the visual damage in this type of fatty embolism is repaired both quickly and spontaneously. The white silvery patches disappear, while the scotoma becomes " perforated " before vanishing. His third case shows the typical ophthalmoscopic picture of the fatty embolism after a soft tissue operation.

We suppose that in these clinical cases, as in our four excised eye specimens, the blood is displaced by the fatty emulsion. This leads clinically to damage and leakiness of the capillary endothelium. Blood and fat escape into retinal tissue.

Surface tension congeals the fatty emulsion in the retinal tissue, and it becomes visible ophthalmoscopically as silver white patches. As there is no other retinal vascular disease, and the retinal tissue is normal, the blood and fatty débris vanish quickly by absorption.

On the other hand, the intra-arterial fatty material is *finely* emulsified, and might pass through the retinal capillaries without damage to the vessel wall and retinal tissue. Both processes might occur simultaneously in traumatic cases.

While the occurrence of fat embolism in a retinal artery of an eye which is to be excised has no practical interest, it may throw light on the mechanics of fatty embolism generally.

We suspect that some of the symptoms ascribed to shock generally may be in fact symptoms of fatty embolism of pulmonary and cerebral capillaries.

Our observation shows the ease with which fatty droplets can enter vessels opened by accidental or operative injury.

This might be even a potential danger in foreign body extraction with the posterior route.

One feature in the causation of traumatic retinal fatty embolism can be understood only with difficulty. As these fatty droplets entering the veins (*e.g.*, of the fatty bone marrow) arrive in the retinal arteries, they are bound to travel through the pulmonary capillary system, yet only some of the traumatic fatty embolism cases show the signs of pulmonary embolism.

Microscopic examination of unstained specimens in our four cases has shown that the intra-arterial fat is very finely emulsified. This fine emulsion might pass the pulmonary capillaries without provoking gross disturbance. We can explain the phenomenon, therefore, without postulating the bypassing of the arterio-venous anastomoses in the pulmonary vascularisation.

Although, so far as we know, no one has previously recorded fatty embolism after excision of the eye, it does not seem to be a rare occurrence. It is clear that routine histological examination would reveal no trace of the intruded fat. We can discover it only after investigation of the fixed and opened eye, preferably by the slit-lamp. Even then, the appearance of the milky bands might be overlooked (as certainly happened to us before we investigated the intra-vascular content) among the collapsed vessels.

We feel sure that systematic ophthalmoscopic investigation of injured patients, especially those with bone fractures (but possibly also without), will show this ophthalmoscopic picture to be frequent. It has escaped our attention as the retinal changes are transient, and do not as a rule leave any diminution of visual function behind.

Summary

Four cases are described in which enucleation or orbital exenteration was followed by fatty embolism of one or more branches of the central retinal artery. The embolism was more or less fluid, and consisted of a fine fatty emulsion which stained shining red with scarlet red. The fatty content of the central artery is assumed to be aspired from the orbital fat and emulsified by the blood. Fatty embolism of the central artery with white silvery retinal patches and superficial haemorrhages is relatively frequent in fractures of the long bones, and is probably the cause of Purtscher's traumatic retinal angiopathy. The proved fine emulsion of the fat in these four cases may explain the so far inexplicable passage of fat through the pulmonary capillaries.

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We are indebted to Mr. Donald for the slit-lamp sketch, and Mr. Watt for photomicros.

ATROPHIA GYRATA CHOROIDEAE ET RETINAE*

BY

JOHAN SÆBØ

NORWAY

IN 1871 Mauthner published the report of an eye-condition, named choroideremia, and in 1896 E. Fuchs described a similar case under the term of atrophia gyrata choroideae et retinae. Later about 35 investigators have reported some 50 similar cases, trying to classify them as belonging to the one or other of these two clinical pictures first described.

It is evident from their information, however, that they have not always succeeded, and on reading the reports one receives an impression of great confusion.

Mainly, however, two divergent conceptions are prevalent, namely:

1. The two clinical pictures described both belong to the same condition, choroideremia forming the ultimate stage in the development of atrophia gyrata.

2. They are seen as different diseases, choroideremia being a defect or malformation, and atrophia gyrata an acquired progressive degeneration (atrophy) of retina and choroid.

Duke-Elder (1946) does not try to conceal the confusion around these conditions, writing: " In view of our unsatisfactory knowledge of both conditions and in the complete absence of pathological studies, the question must be left open; indeed it is questionable whether either condition forms a homogeneous group," and he goes on: " It will also be remembered that no case of choroideremia has been seen younger than 14 years, but at the same time no one has yet seen a case of gyrate atrophy develop into this condition."

Another writer, Bedell (1937) declares: " The cases are too few to warrant dogmatic statements."

The variation in description of different symptoms in these conditions is not so great as to prevent a review of their main common features.

The condition is often familial. Consanguinity has been demonstrated in some cases. The hereditary character of the disease has been mentioned, the recessive modus being specially discussed. But especially on this point the information is unsatisfactory.

Night-blindness is one of the first and commonest symptoms.

* From the University Eye Department, Rikshospitalet, Oslo. Chief: Professor Birger Malling, M.D. Received for publication, January 29, 1948.

Most patients state that their night-blindness has lasted a long as they can remember, others have noticed the symptom first in the twenties. Determination of the commencement of the disease on this basis is therefore difficult.

The actual changes in the fundus oculi have been observed at various ages, down to 10 years (Mori, 1914; Werkle, 1931). Most publications, however, deal with patients of 20 years or more. Central vision becomes affected in an advanced stage, considerably reduced vision or amaurosis then occurring at a relatively early age.

The initial symptoms are usually vague, and this is probably the reason why no one has yet been able to determine just when and how the changes in the fundus take place.

The conditions have in several cases been associated with myopia, sometimes to a considerable degree. The accompanying myopia has even been considered decisive for the diagnosis: choroideremia or atrophia gyrata (Beckershaus, 1926).

The visual fields are in most cases contracted, in a great number of cases to within 10°.

Descriptions of the changes in the fundus vary. No two cases seem alike. In his work Bedell expresses the same opinion when saying: " At the beginning of this discussion I wish to say that practically no two cases are alike in regard to the distribution of pigment, the size and shape of the retained choroid about the macula or the modifications of the retinal and choroidal circulation. Many cases are similar but none are identical."

The changes vary in the different cases as well as in the two eyes of the same patient (Wernicke, 1909). Varying types have also been described within the same family (Böhm, 1932; Werkle).

The changes in the fundus have been described as a more or less extensive defect or atrophy, essentially of the central part of the fundus. The area round the macular region, however, seems to retain its normal appearance for a long time. Pigment and choroidal vessels become reduced and vanish from the atrophic part, which ultimately shows a whitish, glistening field with a few scattered pigment masses. This defect or atrophy may in some cases spread as far as to the periphery of the fundus. In other cases a rim of comparatively normal appearance is seen. This rim, or the remains of it, has been described in choroideremia and in gyrate atrophy.

Most authors claim that normal appearance of the retinal vessels is retained, whereas others, *inter al.* E. Fuchs, that they may be contracted almost to invisibility.

The optic discs have in most cases been described as normal, whereas E. Fuchs and Beckershaus state them to be yellow as in retinitis pigmentosa, and Bencini (1938) to be rose-coloured.

The earlier published records thus offer several common features: familial occurrence, night-blindness, reduction of central vision, defect of the visual field and defect or atrophy of the central part of the fundus, an area around the macular region and a peripheral rim excepted.

The varying descriptions of these conditions in different publications make it difficult to classify them. The characteristics which have been thought to distinguish the two conditions, atrophia gyrata and choroideremia, from one another are not always convincing. The efforts of the various text-books to describe the clinical picture of the two conditions give clear evidence of this difficulty.

The question will then be whether this difficulty can be explained on the basis of the available literature and the present material.

The conditions being rare, few students have had a chance to see many cases. If that should happen, they belong as a rule to the same family and the same generation and are of nearly equal age. It has, therefore, not been possible to observe the progressiveness of the disease. The insidiousness of the initial symptoms prevents an early diagnosis, and the condition has, therefore, never been followed from its beginning. The information given is often somewhat unsatisfactory, and one lacks illustrations of the findings. Therefore, a comparison between the individual cases is often impracticable. There is often no information as to whether the examination has been made in mydriasis and it is just in these cases that it is important to study the peripheral fundus findings in order to make a differential diagnosis between atrophia gyrata and choroideremia.

It has been mentioned that some of the cases are associated with myopia, and it is possible that enough attention has not been paid to the atrophic changes for which the myopic condition itself is responsible.

Further the pigmentation of the fundus is known to vary in light- and dark-complexioned individuals, so that even this factor has to be considered.

Accurate familial investigations are lacking. It is often merely mentioned that no other member of the family is suffering from the disease, or that no consanguinity has been found.

The close connection between these two conditions and retinitis pigmentosa has been a constant source of confusion. Thus Leber (1877) has classified Mauthner's case as an extreme retinitis pigmentosa.

Reports of the examination of the visual fields are seldom supplied with charts.

The presence of night-blindness has as a rule been reported, without any information as to how this has been demonstrated.

Accordingly the publication of every new investigation into these rare conditions seems justified.

First an attempt will be made to give a clinical picture, based on the present material, consisting of a family with 4 affected brothers, aged 27, 38, 44 and 46 years. Next this will be compared with the earlier descriptions to such an extent as is possible.

The present work will possibly contribute to the solution of some of the problems arising in connection with earlier publications with varying titles: choroideremia, atrophy of choroid, retinitis pigmentosa atypica, retino-choroidal atrophy, deficit of choroidea, congenital absence of choroidea with retinitis pigmentosa, congenital deficit of choroidea, etc.

On account of the familial examinations journeys have been made to Solör, the home of most members of the family. Investigations have also been made to the Public Register in Hamar and at the Parish Register.

Dark-adaptation has been examined by means of a biophotometer. The procedure has been: adaptation to dark for 20 min., then adaptation to light for 10 min.

The illustrations of the fundus oculi have been drawn by A. Scheen, Oslo. To include also the peripheral part of fundus, the picture of the central part has been somewhat condensed. The changes in the fundus being decidedly symmetrical in both eyes, the picture of more than one eye from each patient has been considered unnecessary.

Case reports

General examination of the members of the family in question has revealed no other disease nor defect that might be suspected of having a connection with the mentioned eye-condition. All members of the family seemed normally equipped mentally. They were well-built and healthy-looking. Wassermann reaction was negative in the four affected brothers. Neither did the information of causes of death in the family suggest any prevailing familial disease that might be connected with the eye-condition. General examination of the diseased eyes has offered no pathological finding, apart from those in the fundus. The lenses were clear in all the patients. Transpupillary illumination showed a glistening white reflex from the central part of the fundus. Tension was normal in all. All examinations have been made in maximal

mydriasis. The patients were dark-haired and with brownish complexion.

Case 1. I.H., born March 24, 1915, aged 27 years.—The first trouble noted by the patient was difficulty in finding his way home after dark. There was a marked difference between himself and his working companions. He felt inclined to believe this night-blindness to have been present ever since he was a child, but was sure it was there at twenty. He was an average pupil of the preliminary school and reading caused him no trouble. He served his military term as a common soldier. He was working in the woods, and in his work he had no inconvenience from his eyes. He had noticed a slight peculiarity, however, namely that when he was looking for any small object, the location of it took him a considerable time. He had in a way to search for it with his eyes. He had noticed no flickering nor photopsias. Vision, he believed, was still good during day-time, but the diminution of night-vision was troublesome.

Status praesens 5/1/43:

Fundus o. dext. Plate I, Fig. 1.

The optic disc is of normal colour and appearance. Temporally it is surrounded by a narrow white halo. The retinal vessels seem normal. Centrally in the fundus there is a somewhat irregular area measuring about 3 disc-diameters, striking because of its reddish colour in the otherwise whitish annular zone surrounding this area and the optic disc. The appearance of this macular area is practically normal. Temporally it has a somewhat irregular brownish pigment-border with some projections. Otherwise it shows some scattered pigment spots, somewhat more densely accumulated in macula proper. Peripherally there are some traces of choroidal vessels. The borders towards the disc are more diffuse with even transition into an atrophic area with fine pigment granules, specially above and below the disc. Several choroidal vessels are seen in the atrophic part between the disc and the macular region.

This maculo-papillary region is surrounded by a whitish annular zone, varying in width, and peripherally limited by an irregular pigment-border with long pigmented projections into the light annular zone. In turn these may communicate with irregular pigment-masses in the light zone, thus forming lagoon-like designs. Pigment is totally lacking in a great part of this annular zone, and the white sclera is seen shining through. Specially on the nasal side a great part of the choroidal vessels is obliterated, only a few contracted ones remaining in some places. Between the peripheral border in the fundus and the macular area, however, some thick choroidal vessels are seen crossing the white zone.

PLATE I

FIG. 1.

Atrophia gyrata choroideae et retinae. Fundus o.d. Case 1, I.H., aged 27 years.

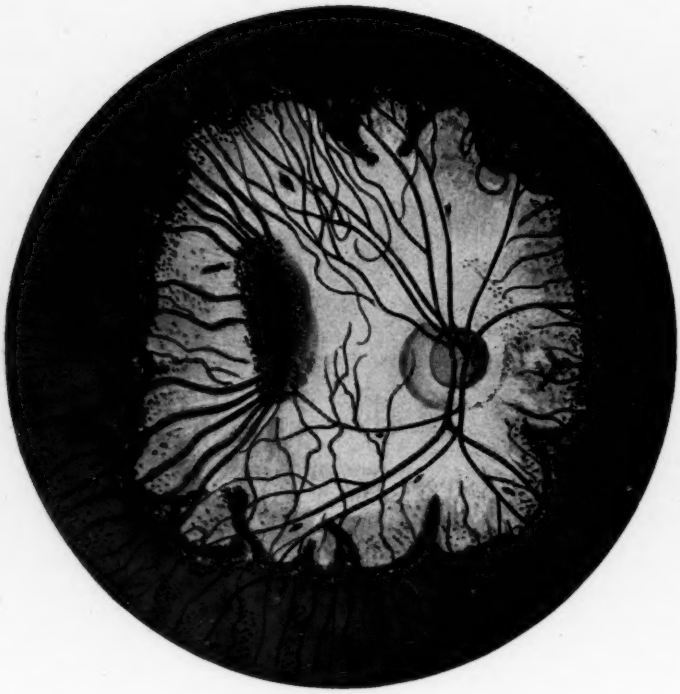

FIG. 2.

Atrophia gyrata choroideae et retinae. Fundus o.d. Case 2, O.H., aged 38 years.

FIG. 3.

Atrophia gyrata choroideae et retinae. Fundus o.d. Case 3, S.H., aged 44 years.

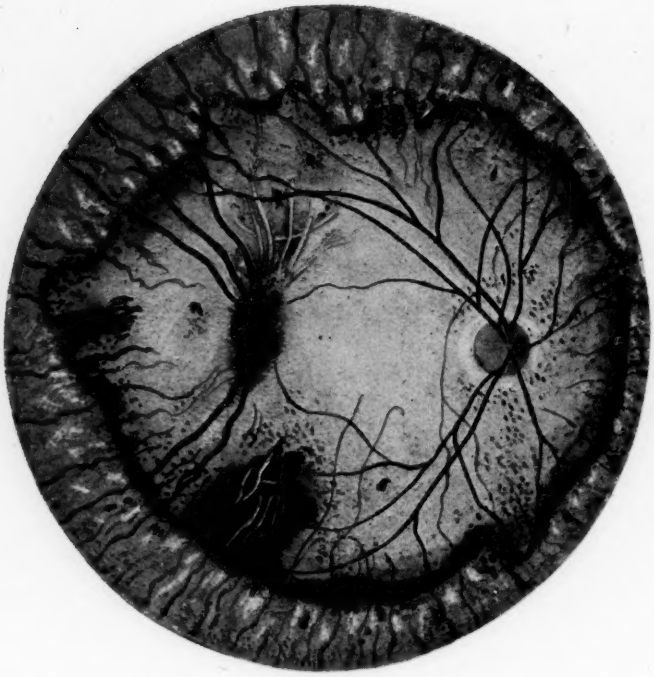

FIG. 4.

Atrophia gyrata choroideae et retinae. Fundus o.d. Case 4, E.H., aged 46 years.

This whitish zone is surrounded by a rim of apparently normal fundus. It is somewhat wider temporally than nasally and centrally it is limited by the above mentioned pigment-border with its projections. The part immediately outside the peripheral border, however, is slightly greyish with irregularities of pigmentation and of atrophic appearance. Near the border the choroidal vessels from the whitish annular zone are distinguishable.

The picture of fundus o. sin. is practically the same. The optic disc and the retinal vessels must be described as normal. The reddish macular area is somewhat larger and more irregular, but otherwise showing the same properties as the right side. Except on the nasal side this area is everywhere surrounded by a pigment-border. The same whitish annular zone is seen surrounding the maculo-papillary region, that peripherally is limited by a pigment-border with projections, showing the same tendency to lagoon-formation. Scattered in the white zone, particularly around the optic disc and between the macular region and the disc, are some larger pigment masses and dust-like dots. The choroidal vessels are highly reduced, a few thick ones still persisting temporally in the zone, specially in connection with upper and lower part of the macular region. The peripheral rim is present, and is of the same appearance as on right side.

Visus : o.d. : 5/5, o.s. : 5/5.

Fields of vision : See Chart, Plate II, Fig. 1.

Sense of colour : Normal according to Ishihara.

Night-vision : See text-fig. 2, p.838. Reduced.

Case 2. O.H., born August 20, 1904, aged 38 years.—The first symptom of eye-trouble noticed by the patient was that his vision in the dark was not as good as that of his friends. He believed night-vision to have been reduced since his early youth, and was certain of it at twenty. He had served his military term, and at that time noticed nothing wrong with his vision. 5 years ago he noticed that his vision had become diminished also in day-time. This had gradually grown worse, so that now he was afraid he would have to give up working in the woods. He was able to read only for a short while at a time and by good light. He had never noticed flickering before his eyes nor photopsias.

Status praesens 7/1/1943 :

Fundus o. dext. See Plate I, Fig. 2.

Colour and appearance of the optic disc are normal. It is surrounded by an atrophic halo. The retinal vessels seem normal. Corresponding to the part around the macular region there is an oval, about 1 x 3 disc-diameters, greyish-red area. Appearance of its temporal part is that of normal fundus, whereas the nasal part

is greyish. Temporally this area has a brownish pigment-border. Corresponding to the macular region proper there is a somewhat denser accumulation of pigment. A bundle of thick choroidal

Plate 11

Visual Fields in Atrophia gyrata Chorioideae et retinae. Obj. 5/33o.

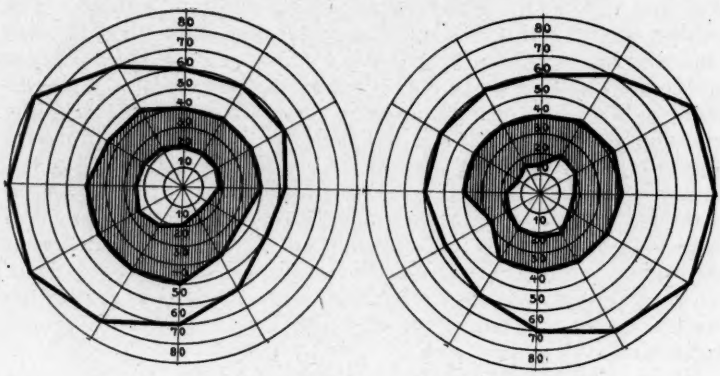

Fig.I.- Case 1, I. H., 27 years.

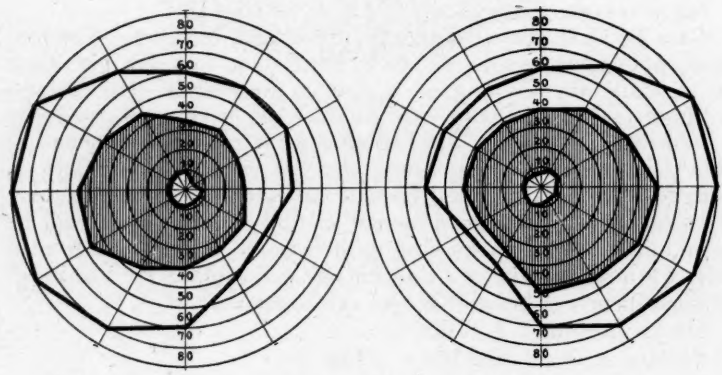

Fig. 2.- Case 2, O. H., 38 years.

vessels lead to the upper and lower pole, and stand out sharply against the whitish sclera. Apart from this maculo-papillary region, the entire central fundus is occupied by a glistening white

annular zone, where, besides the mentioned choroidal vessels to the macular region, only scattered and contracted choroidal vessels and a few smaller or larger pigment-masses are seen. This whitish annular zone is peripherally limited by a border of pigment with irregular projections, capable of forming lagoon-like designs. The peripheral rim in the fundus is present. Close to the pigment-border the fundus contains rich amounts of small pigment-masses, and is somewhat greyish and pale. Otherwise it resembles the normal fundus. Width of the rim is about the same as in Case 1.

Fundus o. sin. The picture of the fundus is practically identical with that of the right eye, making a detailed description superfluous. Disc and retinal vessels appear normal. The macular area has an oval shape and greyish-red colour, with scattered pigment-masses, somewhat denser in the macular region proper, and a more closely-knit bundle of choroidal vessels lead to the upper and lower poles of the region. The central part of fundus is for the rest a glistening white with very few and contracted choroidal vessels. There are scattered masses of pigment. The zone is peripherally limited by a similar pigment-border as on the right side. The peripheral rim in the fundus is somewhat greyish and atrophic with some pigment-masses near the border, but otherwise apparently normal.

Visus: o.d.: 5/50, o.s.: Counting fingers at 3 m.
0° + 0·5 ÷ 3 5/10; 0° + 1 ÷ 3 5/50.

Fields of vision: See Chart, Plate II, Fig. 2.

Sense of colour: Normal according to Ishihara.

Night vision: See Text-fig. 2, p. 838, Highly reduced.

Case 3. S.H., born July 19, 1898, aged 44 years.

The first eye-trouble noted by the patient was difficulty in seeing in the dark. He was unable to determine the time when this began nearer than to early youth. He had served his military term as a common soldier. At thirty-seven he knew for certain that his vision had become diminished also in day-light. Since then his eye-sight has steadily diminished. He worked in the woods, and his eye-sight was now interfering with his work. He had been an active hunter, but had to give it up 5 years ago. He could read big print only. He had never noticed flickering before his eyes nor photopsias.

Status praesens 5/1/43:

Fundus oculi dext. See Plate 1, Fig. 3.

Colour and appearance of the optic disc are normal. It is surrounded by a peripapillary halo. The retinal vessels are of normal appearance. Corresponding to the part around the macular region there is a triangular, faintly greyish area. By a projection

its temporal corner is connected with the peripheral rim of the fundus. This area has a slightly pigmented border, and scattered over the triangular space there are fine pigment-masses with a denser accumulation corresponding to the macula proper. The macular region is distinctly atrophic, the underlying choroidal vessels faintly visible. Some choroidal vessels enter this region from the periphery, especially to the upper and lower part. Below this triangular space there is a larger dark brown pigment-mass with several sharply outlined round holes, through which the white sclera is visible. At the bottom of these holes a few choroidal vessels are seen. Outside these areas the central fundus is occupied by partly glistening whitish areas with no or few and contracted choroidal vessels. Scattered over this whitish annular zone are small pigmented masses or greyish areas. Also some larger pigment-masses are seen, partly communicating mutually, partly with the peripheral pigment-border, thus forming lagoon-like designs. The peripheral rim, present also here, is centrally defined by a distinct pigment-border. Peripheral to the pigment-border the rim is greyish-brown with scattered pigment-masses and is distinctly atrophic. At the extreme periphery the rim is of a brighter reddish colour, but even here it cannot be described as normal.

Fundus o. sin. The picture seen here is mainly the same as on right side. The macula area is somewhat larger and more irregular with pigmented projections communicating with the pigment-border of the peripheral brim. The macular area is surrounded by a pigment-border and has a greyish-red colour and fine pigmented spots, densest in the macular region proper. A bundle of thick choroidal vessels is seen leading to the upper and lower parts of the macular region. Otherwise there is a similar central annular zone with glistening white parts and pigment-masses with lagoon-forming projections. The peripheral rim in fundus is like that in right eye.

Visus: o.d. 5/20, o.s. 5/50.

Barely passing Schiötz reading test 1·20 with right eye.

Fields of vision: See Chart, Plate III, Fig. 3.

Sense of colour: Apparently good according to Ishihara.

Night-vision: See Text-fig. 2, p. 838. Highly reduced.

Case 4. E.H., born December 22, 1895, aged 46 years.

In early youth already the patient noted that his vision after dark was not so good as that of his friends, but he was unable to fix the time for the beginning. At twenty his vision was reduced also in day-light, and at thirty-five he was no longer able to read. Since the age of forty he has been blind. He has never noticed flickering before his eyes nor photopsias.

Plate III

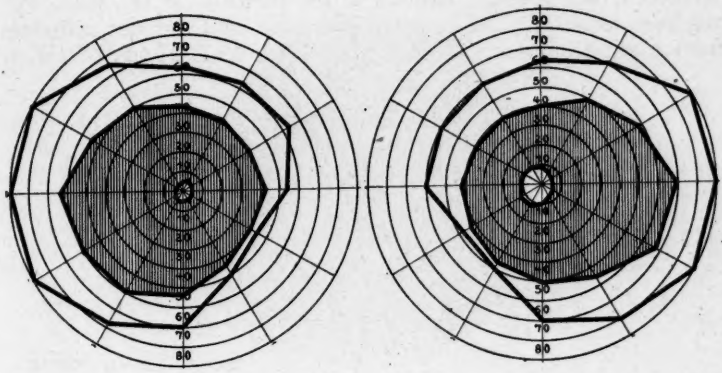

Fig. 3.- Case 3, S. H., 44 years.

Status praesens 5/2/43 :

Fundus oc. dext. Colour and appearance of the optic disc are normal. It is surrounded by an atrophic halo. The retinal vessels are possibly somewhat contracted. A greyish-brown, about disc-sized area is here seen around the macular region, with a few choroidal vessels to the upper and lower pole. Above and below this macular area there are some greyish-brown parts with some lighter streaks and lines, interpreted as remnants of atrophied choroidal vessels. The macular region is changed, having lost all its normal structure. Otherwise the entire central part of fundus is whitish and glistening, with scattered pigment-dots and a few larger masses, particularly in the vicinity of the optic disc. The choroidal vessels are mostly obliterated, leaving a few contracted ones in the extreme periphery of the white zone. The peripheral rim is present, but narrower than in the other patients, and more greyish-pale, atrophic. Centrally it is limited by a pigment-border, somewhat less prominent than in the others. The pigmented, lagoon-forming projections are seen also here.

Fundus o.s. The picture being identical to that of right eye; further description is considered unnecessary.

Visus : Reduced to mere perception of light o.u.

Family investigations (See pedigree, Plate IV)

The disease is demonstrated in 4 brothers in a family of 9 children, 6 men and 3 women. The brothers and sisters of the patients and all their children have been examined and found normal. The only child of one of the patients (2, O.H.), a boy of seventeen, was normal. Mother of the patients (M.H., Gen. IV) has been examined. She was 70 years old, and was not suffering from night-blindness. Central vision 5/5 o.u. Fields of vision

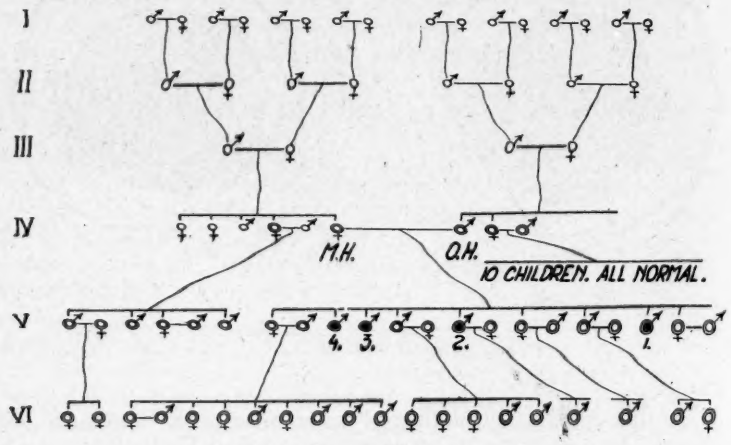

PLATE IV.

Pedigree demonstrating the family with four cases of atrophia gyrata choroideae et retinae.

◎ examined. ◉ affected.

were normal. The optic discs were surrounded by white atrophic haloes with a few choroidal vessels (see Text-fig. 1). This finding was read as a senile atrophy, and is specially mentioned here because similar findings have been described in literature (Bencini), and it will presently be discussed.

Father of the patients (O.H., Gen. IV) was 69 years old and had normal eyes. His sister (O.F., 64 years old) and her 10 children have all been examined and their eyes found normal. M.H., Gen. IV, was one of a family of 5, of whom 3 are dead at 64, 25 and 26 years of age. None of these were known to have had eye-trouble. The surviving sister and all her descendants were normal.

The direct line of ascendants of M.H. and O.H. has been followed for 3 generations, information being sought in the parish

and public registers and from the living members of the family, without evidence of eye-diseases in any of these individuals being found. Neither has consanguinity of M.H. and O.H.'s families been demonstrated. The possibility of existing blood-relationship cannot be excluded, however, on basis of these investigations,

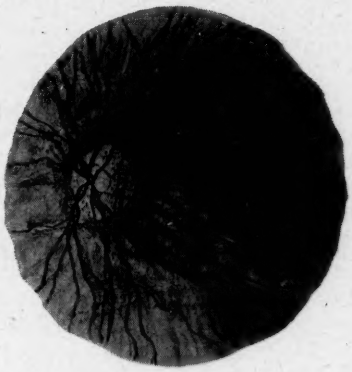

FIG. 1.

Atrophic halo around the optic disc. M.H., mother of the patients (drawing by the author).

specially including ascendants in direct line. But all members of the family denied the existence of such connection between the two families. M.H. and O.H. came from communities far apart.

Thus the disease has been demonstrated in 4 brothers, but neither in their ascendants nor descendants.

Clinical picture

Histories and clinical pictures of the 4 brothers are strikingly uniform. The first symptom of the condition is night-blindness. The patients are unable to fix the time for its beginning, but as a rule state that it has been present since early youth. The youngest patient (Case 1. I.H., aged 27 years) declares that his night-blindness probably commenced when he was a child, but he was sure of its presence at twenty. Table I below gives the time when night-blindness and diminished central vision were noticed by the patients, and vision at the time of examination.

The table gives evidence that night-blindness is the initial symptom, being noticed by the patient before the reduction of central vision. Thus the 27 years old I.H. has still normal vision. The other patients have noticed diminished vision at 33 and 37 and

TABLE I

Age at time of examination—beginning of night-blindness and reduced vision—central vision in atrophia gyrata choroideae et retinae.

Case	Age	Patients noticed :—		Central vision
		Night-bl.	Red vision	
1.	I. H., 27 years	In childh.	Gives normal vision	0.d. : 5/5, o.s. : 5/5
2.	O. H., 38 years	At twenty	At thirty-three	0.d. : 0°+0·5÷3 : 5/10 0.s. : 0°+1÷3 : 5/50
3.	S. H., 44 years	In youth	At thirty-seven	0.d. : 5/20, o.s. : 5/50
4.	E. H., 46 years	In youth	At twenty	Amaurotic at forty

20 years. In case 2, O.H., diminished vision has been demonstrated objectively at 38 years, and in case 3, S.H., aged 44 years. He is now barely able to read big print, and is having trouble with his eyes when working in the woods. Case 4, E.H., aged 46 years, has been blind since the age of 40 years.

None of the patients has noticed flickering before his eyes nor photopsias.

Determination of the time for onset of the disease must essentially be based on the patients' statement that night-blindness commenced in childhood or early youth.

The actual changes in the fundus oculi show great uniformity in the 4 patients, the only variations demonstrable being supposedly due to progress of the disease. The changes are also mainly symmetrical for both eyes.

The optic discs are of normal appearance. So are the retinal vessels, Case 4 excepted. This patient is blind, and the retinal vessels are here somewhat contracted. The macular region seems to remain normal for a long time, presenting the picture of a more or less reddish island in the otherwise whitish and glistening central fundus. This macular area is in most cases surrounded by an irregular brown pigmented border. Within the area there are scattered pigment-dots, often somewhat denser in macula proper. With age it becomes concentrically reduced, gradually assuming a more greyish-brown colour, until, as in the 46 years old patient (Case 4, E.H.) merely an irregular greyish-brown patch remains.

In connection with the macular area there are some choroidal vessels, specially around the upper and lower part, and from here crossing the whitish atrophic zone toward the periphery. Number and calibre of these vessels decrease with age of the patient. In the eldest brother (see Plate I, Fig. 4) only a few thin ones remain. Near the macular region in this patient there are some whitish lines in a denser accumulation of pigment-masses. These lines are undoubtedly attributable to obliterated choroidal vessels.

The central part of fundus, apart from this macular area, is essentially changed into a more or less glistening, white annular zone, peripherally limited by a narrow retino-choroidal rim. Besides the above mentioned choroidal vessels, the younger patients show, specially in the macular region, also a few scattered ones in the whitish annular zone. These vessels become reduced with advanced age, until they show nearly complete obliteration in the oldest patient. Scattered pigment-dots and masses are found also in this zone. A number of pigmented projections from the peripheral rim show a tendency to lagoon-formation. The position of the pigment-masses and projections suggest an earlier connection between these. The described pigment formations present a highly irregular picture, different from the more regular " bone-corpuscle cells," of retinitis pigmentosa. Also the pigment-accumulations seem to become reduced with advance of the patient's age.

The retino-choroidal atrophic zone widens in the course of years, essentially through contraction of the macular area, but also at the expense of the peripheral rim.

This retino-choroidal rim is seen in the extreme periphery. In the younger patients the rim, close to the border, is somewhat greyish-atrophic with a few dots of pigment. Otherwise it is normal. In the oldest patients it is possibly all atrophied. Width of the rim is also somewhat reduced in the oldest patients. An estimation of this part, however, is admittedly difficult, even in good mydriasis.

Transpupillary illumination, as it has been mentioned, gives a white reflex corresponding to the annular zone in the fundus. The lenses are clear in the 4 patients.

Sense of colour is normal.

Adaptation to dark is greatly reduced. (See Fig, 2, demonstrating curves for the 3 examined cases, and a normal curve.)

The fields of vision show characteristic changes in accordance with the changes demonstrated in the fundus oculi. The defects of the visual fields are mainly symmetrical in the two eyes (see Plates I and II). The peripheral parts are normal, but all cases show

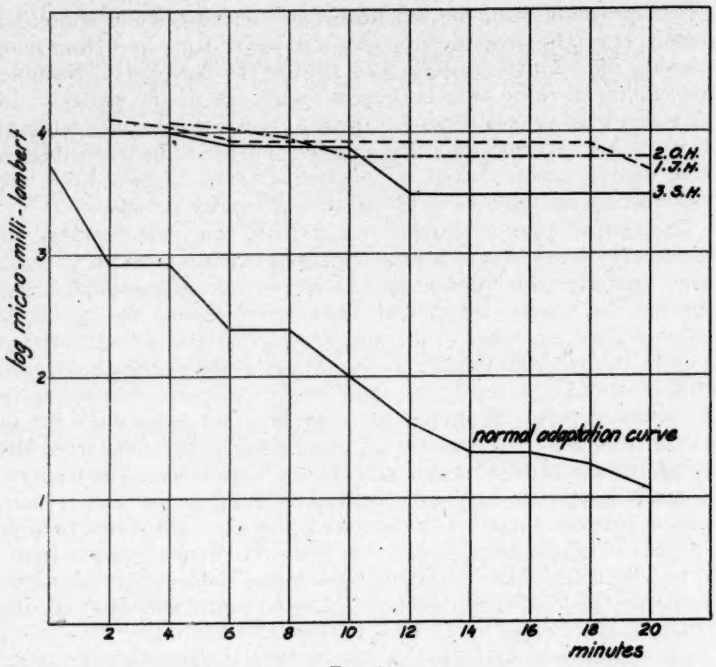

FIG. 2.

Atrophia gyrata. Curves for dark-adaptation. Biophotometer. Adaptation to dark for 20 min., followed by 10 min. exposure to light before examination.

almost absolute annular scotomas. These widen with the advance of age, partly through peripheral spreading, but essentially through contraction of the central parts. In S.H., aged 44 years, they remain only an area inside of 10°.

The condition is familial, being demonstrated in 4 brothers of a family of 9 children. It has neither been traced to their ascendants nor descendants. Consanguinity of the parents has not been demonstrated here. The condition is probably due to a recessive gene, a possibility that will be more closely discussed in the following.

Discussion

The picture presented by the 4 patients is unquestionably that of a clinically uniform and progressive chorio-retinal condition, answering the diagnosis of atrophia gyrata choroideae et retinae. Discussion of the differential-diagnosis with regard to the various

types of retinitis pigmentosa, retinitis albescens, coloboma retinae, etc., has been considered superfluous.

As already mentioned, the finding by several authors (Jacobsohn 1888, Beckershaus, Böhm, Smith and Usher 1916, and Zorn 1919), of retinitis pigmentosa within families where chorioideremia and gyrate atrophy have been demonstrated, has caused much confusion around these conditions. If so, it may be questioned whether the latter conditions and retinitis pigmentosa occur independently, or whether there exists a more intimate connection between them. This question is difficult to answer on account of the often unsatisfactory information given in descriptions of the cases. Jacobsohn has thus named his case "retinitis pigmentosa atypica," Leber (1877) has characterised Mauthner's case as an extreme retinitis pigmentosa, whereas Nettleship (1908) considered a connection between these conditions and retinitis pigmentosa improbable.

Choroideremia is the condition most commonly described in connection with retinitis pigmentosa. Somewhat strange, however, seems Beckershaus' finding of a yellowish-white optic disc and "fadendünn" retinal vessels in this 33 years old patient in a family where retinitis pigmentosa had been demonstrated. This also applies to Böhm's case, in which the optic disc was pale and the retinal vessels contracted.

It will be emphasised in this connection that no case of retinitis pigmentosa has been found in this examined family, where the clinical picture is unmistakable.

Beginning of the disease. Determination of when and how the disease has commenced, has not been possible on basis of the present material. The appearance of night-blindness proves that it begins in early youth. Usher (1935) states the age of his youngest choroideremia patient to be 14 years. Average age of the patients at the time of the diagnosis was 27.45 years for choroideremia, and 17.36 years for gyrate atrophy. Werklē has shown gyrate atrophy in a patient aged 10 years. When and how the condition has begun has therefore never been stated. Thus these factors offer no certain clue to the recognition of various types of the disease.

In the present cases the condition has been followed from the age of 27 to 46 years, and a steady progression has been observed. Even in the 27 years old patient (Case 1, I.H.) considerable changes were demonstrated in the fundus, in spite of the relatively inconsiderable subjective symptoms. Judging by this development, the first beginning must have taken place very early in life.

The changes in the fundus oculi are mainly symmetrical in the

two eyes, and similar in the 4 patients. The demonstrated variations must be attributed to progress of the condition. Comparing the changes in the fundus in the present material to earlier descriptions is rather difficult, because of the often unsatisfactory information given. Bedell's statement, quoted in the introduction, may seem justified. A number of reports show accordance with findings in the present cases, but others show great variations.

It must be kept clearly in mind, however, that the question is of a homochronous condition with a progressive course, and that other factors, *e.g.*, excessive myopia and the patient's complexion may influence the phenotype of the disease. Also other ocular conditions and eventual diseases may partly be traced in the picture.

The atrophic changes in the present cases are localised to a certain part of the fundus. This cannot be expected to be alike for all cases, and evidence of variations on this point is found in the literature. Thus McGuire (1932) in an 11 years old patient with atrophia gyrata, has found atrophy of the extreme part of fundus. Cutler (1895) found an atrophic zone around the optic disc, then a normal annular zone and more peripherally another atrophic zone. (Cutler's patients are the same ones that have been described by E. Fuchs.) In such cases the total clinical picture will be decisive for the diagnosis.

In the present cases there has been found a more or less normal looking chorio-retinal rim. This finding varies in the literature and it can therefore not be considered characteristic for those cases considered as atrophia gyrata. This rim, or parts of it, has also been mentioned in cases described as choroideremia or total atrophy. (Alexander 1910, Connor 1919, Zorn, Wolf (1930), Parker and Fralick 1931, Smith and Usher 1916, and Bedell). Neither does this point seem to make a difference between the two described types, which is also in accordance with the findings in the present material. Continued atrophy in the oldest patient is here believed capable of causing complete disappearance of the rim, thus at this stage justifying the diagnosis of choroideremia. In this patient the peripheral rim was already at forty-six hardly detectable even in good mydriasis.

Choroideremia having been described as a defect or malformation, progress of the condition might *a priori* be thought particularly prominent in gyrate atrophy. Not even this factor makes a distinction between the two types possible. Alexander has thus observed progressiveness in " congenital absence of the choroid." Usher and Smith (1916) have described choroideremia in a 24 years old patient. On re-examination 19 years later there was found

striking progress of the condition. Usher (1935) himself says that gyrate atrophy would probably have been found on examination at an earlier stage. Also the finding on first examination of " dark red areas at the extreme periphery of the fundus " is worthy of notice.

Wernicke has observed definite progressive changes in a case of atrophia gyrata. He further found a picture resembling that of atrophia gyrata in the right eye and of choroideremia in left eye. Werkle has studied the condition in two brothers, aged 10 and 20 years, and in another patient aged 50 years, and does not disregard the possibility of the two types representing different stages of an identical disease. Neither has the study of earlier descriptions nor of the present material revealed peculiarities in the changes of the fundus capable of characterising the two types.

Normal retinal vessels and optic discs have been demonstrated in most descriptions of both types, whereas changes have been mentioned in some cases. Reference is here made to what previously has been written (*v.s.*), and only the finding in the present cases of normal discs and retinal vessels recalled. The latter have possibly been somewhat thinner than normal in the oldest patient. In patients where retinitis pigmentosa and choroideremia with papillary and vascular changes have been found in the same family, reliability of the diagnosis may possibly be questioned.

Refraction. Myopia. Beckershaus believes myopia to form part of the picture of atrophia gyrata, using this factor as differential-diagnosis with regard to choroideremia.

Excessive myopia was present in the cases first described (E. Fuchs). Usher has in his studies of the earlier publications often found mention of myopia, also in choroideremia. In this condition myopia was found in 35 of 39 eyes, in 23 thereof not exceeding 3 D. Maximal myopia was −16 and −13 D. In atrophia gyrata 23 of 35 eyes were myopic, and in 17 of these the myopia exceeded 3 D. Maximal myopia was −18 and −20 D.

Myopia has been demonstrated in one case only of the present material (Case 2, O.H.), and here to a moderate degree. Accordingly the symptom of myopia can hardly be used for differentiation between the two types. On the other hand myopia may, *qua* such, with the consequent atrophic changes partly influence the clinical picture.

Fields of Vision. Few publications are supplied with charts. As a rule there is merely a mention of concentric contractions. From the literature Usher has found the following for choroidermia.: Fields of vision are concentrically contracted in 19 cases of 22, are full in 1 case, and in 2 there are no records; ring

scotoma in 3 cases. For atrophia gyrata he found: Fields of vision were full in 5 cases of 26, concentrically contracted in 16 cases, no note in 5, ring scotoma in 1.

The finding of more cases of ring-scotoma in choroideremia than in atrophia gyrata is somewhat surprising. The opposite result might be expected.

Typical annular scotoma is found in all our cases, corresponding to the changes in fundus. The fact that annular scotoma in certain cases is easily overlooked, may be a possible explanation of this disagreement.

Also in Alexander's case of choroideremia annular scotoma has been shown, but this diagnosis was, as mentioned, possibly not quite reliable.

Obviously the fields of vision will change with development of the condition. Annular scotomata will be present in some cases, but are hardly capable of characterising various types of these conditions.

Central vision. The above Table I (p. 836) shows the reduction of vision to be considerable, and capable of causing blindness at forty. Long before this, however, the patients are greatly inconvenienced by the accompanying night-blindness and the reduced fields of vision. From the literature the reduction of vision is seen to be considerable in most of the reported cases. For choroideremia Usher thus finds: Of 34 eyes, vision below 6/60 in 9: for atrophia gyrata, of 35 eyes, vision was below 6/60 in 9. For both types the reports were: Vision ranged from 60/60 to p.l.

Sense of colour is normal in all the examined individuals, in accordance with reports from literature.

Night-blindness. The 4 affected persons have complained of night-blindness. This symptom is also constantly found in the earlier descriptions. Objective examination with biophotometer (see Text-fig. 2, p. 838) shows dark-adaptation to be greatly reduced in all patients.

Lenticular opacities. As already mentioned, all lenses were clear in the present cases of atrophia gyrata. Waardenburg (1939) claims posterior cataract to be a usual occurrence in atrophia gyrata, but rare in choroideremia. Usher reports opacities in the lenses in 29·41 per cent. of his choroideremia cases and in 58·6 per cent. of atrophia gyrata cases. Also this factor is of little use for the diagnosis of the different types.

Aetiology and pathogenesis. The condition is now known to be hereditary and thus connected with the genes. The description of the one type as a congenital defect or malformation and

the other one as an acquired atrophy is therefore incorrect. Both are congenital, but their presentation may vary. This is common in the so-called homochronous conditions.

As for the pathogenesis of the conditions it is only known that a gradual atrophy of choroid and retina takes place in the central part of the fundus. The choroidal vessels diminish and vanish, the pigment disappears essentially from the atrophic parts, being deposited along the borders as the process progresses. The starting point for the atrophy is unknown, but a glistening, white annular zone will gradually develop around the macular region and the optic disc. The process extends somewhat peripherally, but essentially it spreads through concentric contraction of the macular area. The appearance of the macular region remaining normal for so long a time, is explainable by an independent blood-supply of this region. The pigment-projections with their lagoon-like formations indicate a progressive process. In the eldest of the affected brothers (see Fig. 4, Plate I) distinct traces are seen of former choroidal vessels.

No pathologic-anatomical examination of this condition has been made.

Family investigations. Heredity. It has already been mentioned (p. 834), that family examinations including 6 generations, indicate a recessive mode of heredity.

Usher has collected the earlier results in this field, presented below in Table 2.

TABLE II

Demonstrating manifestations of choroideremia and atrophia gyrata within the families. (Usher, 1935).

Type	Number of cases	Number		Number of families	Consang. in No. of families	Appearance of the affected families in No. of generations
		M.	F.			
Choroideremia	22	21	1	19	0	All in one generation
Atroph. gyrata	26	15	11	14	5	4 in 1 gen., 1 in 3 gen. 1 in 2 generations

Of 22 patients with choroideremia only 1 was female. They were all from one generation. 26 atrophia gyrata patients, distributed among 14 families, showed a female contribution of 11. In 4 families the patients were all from one generation, in 1 they are found in 3 generations (Werkle), and in 1 family in 2 generations (Jacobsohn).

In no case of choroideremia has consanguinity been demonstrated, whereas such relationship has been found in 5 of 14 families with atrophia gyrata (Cutler, Mori, Komoto, 1914; Böhm and Arganaraz, 1917).

The finding of consanguinity in 5 of 14 families with atrophia gyrata strongly indicates a recessive mode of heredity.

Where the condition has been traced through 2 and 3 generations, the information obtained is too scarce for further estimation. Werkle's work includes 13 night-blind individuals in 3 generations. Only 3 of these patients have been more closely described, and only 6 of 13 examined. The same uncertainty attaches to Jacobsohn's case. Manifestation of the condition in several generations is therefore not considered definitely proved.

The present material consists of 4 male patients. The above table shows choroideremia to be found mostly in males, while atrophia gyrata is more evenly distributed among the sexes. The possibility of gonosomal heredity suggests itself. However, the available information concerning the affected women's families is too vague for solution of this problem.

Benzini (*v.s.*) noticed an atrophic halo around the optic disc in the mother of one of his patients with choroidal atrophy, and therefore suggested that the condition might be dominant. As already mentioned similar findings have been made in the present material, but the atrophic halo has been considered a senile phenomenon.

Waardenburg (1939) reported a case of atrophia gyrata choroideae et retinae and stated:

1. Consanguinity in the families is found in 40 per cent. of atrophia gyrata and never in choroideremia.
2. Cataracta posterior is common in atrophia gyrata, but is very seldom in choroideremia.
3. In atrophia gyrata females are often affected, but in choroideremia only in 1 of 22 cases.

It seems evident from what is said in the above discussion and from the statements in the present material that these signs are not reliable in differentiating between distinct types of this disease.

Therapy. No effective method for dealing with the condition itself is known. But the patients' families should be told of the eugenic measures for preventing further distribution of the condition. They should then be informed of the laws of heredity for recessive conditions. Preferably, neither the patients themselves, nor their brothers and sisters should have any children. If any of them should wish to marry, he or she should be told of the danger of intermarriage.

In the present cases the patients themselves have seen the responsibility conveyed by the heredity of this serious disease, and have accepted its consequences. Case 2, O.H., was married and had a child already at the time of examination. This latter (aged 17 years) was normal on the last examination. The other patients have remained unmarried.

Conclusion

Homogeneity of the clinical picture and progress of the disease in the 4 affected brothers have been shown. The progress of the process in these patients is so marked that on further development it must be presumed to result in total atrophy of choroid and retina. The condition has been established at an age of 27, but considering its appearance at this age, compared to the development observable in the patient of 46 years, its beginning at a very early age seems probable. This also agrees with the hereditary nature of the condition.

The clinical picture presented has been found corresponding to the term atrophia gyrata choroideae et retinae, the annular atrophy in the central part of fundus being its most conspicuous symptom.

One has compared this clinical picture with those described in the literature as far as this has been possible on account of inadequacy of earlier descriptions. Comparison of the symptoms, one by one, has revealed no certain evidence of these conditions representing two different types: choroideremia or atrophia gyrata.

Here the clinical picture of the disease atrophia gyrata is given, and until a distinct clinical picture of choroideremia, different from this is stated, it can not be justified to continue regarding them as different diseases.

Summary

The present work gives the clinical description of atrophia gyrata choroideae et retinae in 4 brothers.

First a review is given of earlier problems in connection with this condition. Earlier literature has specially discussed whether atrophia gyrata and a condition described as choroideremia should be regarded as two different diseases, or whether they merely represent two developmental stages of an identical disease. Reference is here particularly made to statements by Duke-Elder and Bedell, clearly showing the clinical picture of the two conditions to be far from clarified.

A number of reports, possibly responsible for this uncertainty, are pointed out.

Next follows a clinical description of 4 male patients belonging to the same generation (27, 38, 44 and 46 years old), the clinical

picture of whom is seen to answer to the description atrophia gyrata choroideae et retinae. There are further coloured illustrations of the changes in fundus oculi, charts showing the fields of vision, a diagram showing the results of biophotometrical examinations of dark-adaptation and there are family investigations with a pedigree.

The examinations show a uniform clinical picture in the 4 brothers, the only demonstrable variations being attributable to progress of the condition with the advance of age. Considering the development of the condition in these patients from an age of 27 to 46 years, its first beginning at an early age seems probable. It seems reasonable to presume that continued development of the condition would result in total atrophy of choroid and retina causing the picture to resemble that of choroideremia. On basis of the present cases, however, the annular atrophy in the central part of fundus has been found to be the most prominent symptom, and the clinical picture answering the term of atrophia gyrata choroideae et retinae.

In an attempt to establish symptoms capable of characterising different types of these conditions, the clinical descriptions of the 4 patients and their symptoms have been compared, one by one, to those found in the literature. The comparison has been complicated by the often somewhat unsatisfactory descriptions. However, nothing has been found to justify the continued classification of the two conditions: atrophia gyrata and choroideremia, into two different types. This does away with the earlier conception, mentioned in the introduction, of the former being an acquired, progressive degeneration (atrophy) and the latter a chorio-retinal defect or malformation.

This is also in accordance with the fact that a division into different types has not been possible on basis of heredity, both types apparently being due to recessive genes.

Finally the eugenic measures have been mentioned, that ought to be taken to prevent further distribution of the condition.

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INTRA-OCULAR PHAKOMATA—A REPORT OF THREE CASES*

BY

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IN 1932 van der Hoeve grouped the syndromes of Bourneville, of von Hippel and Lindau, and of von Recklinghausen under the title of the "phakomatoses." Later the syndrome of Sturge-Weber was added, making a fourth.

* From the Institute of Ophthalmology, University of London.
Received for publication, April 24, 1948.

He called the tumefactions found in these diseases " phakomata," from " phakos," the Greek name for mother spot. He described by the word " phakos," a spot, congenital in origin, often hereditary and familial, and which can be found in different parts of the body. It can be present at birth or appear later on, can vary in size, enlarge by proliferation of any part of the tissue, grow to real blastomata and even turn to malignancy. A phakos may be present in any part of the human body.

All the phakomatoses have an hereditary character, although direct evidence of this may not be discovered in many families. The full syndromes are often not seen in all members of a family and mental changes may be completely absent. The discovery of retinal phakomata may be most important in clinching an otherwise doubtful diagnosis. The three patients described below have very obvious stigmata, but all show interesting ocular features that have been considered worthy of record.

Two are cases of phakomata-Bourneville (tuberose sclerosis, epiloia) and one of phakomata-von Recklinghausen (neuro-fibromatosis). The patients were low grade idiots with epilepsy. Each showed cutaneous and retinal spots (phakomata). Intracranial lesions must have been present in addition.

In the family histories no similar conditions are recorded, but the relatives could not be investigated for incomplete manifestations of the syndromes.

Case Reports

CASE NO. 1. E. H. K. Female. Born 1908, died 1936. In 1932 when admitted to mental hospital she was stubborn and resistive. Her physique was poor and she had a left-sided hemiplegia. Adenoma sebaceum was present on the face over the butterfly area and chin. There were some fine spider naevi among the nodules. The pupils were noted as equal and reacting to light and a routine photograph showed left convergent strabismus (Fig. 1). Major epileptic fits numbered about a dozen per annum.

In January, 1933, both pupils were noted as slightly enlarged, the right more than the left, and reacting sluggishly to light.

In September, 1933, the patient's vision appeared to have deteriorated. Early bilateral papilloedema was found with the retinal spot (Fig. 3). A month later a right ptosis and weakness of right internal rectus muscle developed (Fig. 2).

During 1934 the patient became stuporose, incoherent in speech and appeared oblivious to her surroundings. At times she cried out with pains in the head.

FIG. 1.

Case No. 1 showing adenoma sebaceum, left internal strabismus.

FIG. 2.

Case No. 1 showing adenoma sebaceum with right ptosis and weakness of right internal rectus muscle.

During 1935 the pupils became dilated with no reaction to light. Both discs were dead white with very small retinal vessels (secondary optic atrophy). Coarse nystagmus was present. The ptosis and strabismus persisted. During the year she showed signs of further increase in intracranial pressure, the fits became more frequent and she lived a "vegetable existence" until death in December, 1936.

Post-mortem showed numerous tuberous masses in the cerebral hemispheres, particularly in the temporal lobes. There was a large spongioblastoma within the third ventricle with advanced internal hydrocephalus.

Fig. 3 shows the right fundus when papilloedema was present. Just above and temporal to the disc, surrounded by the branching superior temporal artery, there is a sharply defined, circular, flat mass within the retina. Its size is slightly larger than the optic disc. Its colour is chalky white with streaks of yellow and its surface is rough and coarsely pitted. It is surrounded by a broad slate-grey matte ring that gradually fades into the surrounding retina with slight radial extension towards the optic disc. It is a typical retinal phakos as found in Bourneville's disease.

The case is interesting in showing the syndrome of epiloia with retinal phakos, and ventricular tumour causing internal hydrocephalus and raised intracranial pressure leading to ocular palsy and secondary optic atrophy.

CASE NO. 2. F. S. Female. Born 1900, still alive aged 48 years. She is fifth in a mixed family of six; the others are said to be normal. She is unstable, destructive, impetuous and resistive unless managed very quietly and patiently. She is unable to do any of the routine mental tests. Epileptic fits number six to twenty per annum.

She shows very pronounced adenoma sebaceum of the face, particularly on the naso-labial folds and chin. There are some smaller nodules on the upper lip and fewer of varying size on the forehead (Fig. 4). Fine spider naevi are present in association with the nodules.

Ocular examination. No abnormality has been detected in the right eye. Except for the phakomata, the left fundus (Fig. 5) is normal. Below and temporalwards from the left disc, surrounded by branches of the inferior temporal vein, are two chalky-white masses close together within the retina. Each is semi-circular, in size about one quarter of a disc diameter. Their surfaces are flat and finely rough. They are surrounded by a slate-grey area that gradually fades into the surrounding retina. They are retinal phakomata of Bourneville's disease. No change has been noted for many years.

FIG. 3.

Case No 1. Tuberose sclerosis—retinal phakos, papilloedema.

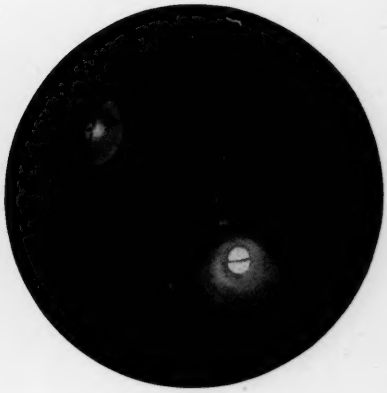

FIG. 5.

Case No. 2. Tuberose sclerosis—retinal phakomata.

FIG. 7.

Case No. 3. Neurofibromatosis of choroid, circulatory disturbances around macula, secondary optic atrophy.

FIG. 4.

Case No. 2 showing extensive adenoma sebaceum of case.

FIG. 6.

Case No. 3 showing small pigmented spots on forehead. Subcutaneous neurofibromata are not seen in photograph.

The patient is interesting in showing the above features and for having lived so long despite her handicaps. For several years she has had, in addition, active pulmonary tuberculosis. This has slowly progressed to become bilateral, but has elicited a very good fibrotic response and her general physique is good. There is no evidence of raised intracranial pressure.

CASE NO. 3. H. C. Male. Born 1898, still alive aged 50 years (Fig. 6). His mental deficiency was noted when he was aged 5 years. To his family's knowledge he is the only mental case for four generations.

On admission in 1931, he was found to be a low grade imbecile who spoke little and replied mostly by signs. At first he had infrequent attacks of petit mal, but over several years these increased to fifty to eighty per annum with some grand mal attacks.

His skin shows several patches of brown pigmentation, particularly on the abdomen, with a few on the face. Multiple subcutaneous fibromata are scattered over the entire body. Numerous venous varicosities are present in each groin and in other sites near the fibromata. The third or fourth lumbar spine appears deficient (possible spina bifida occulta) and both knee and ankle reflexes are absent. His general condition has changed little since admission.

Ocular examination. Tension by palpation appears normal. Both optic discs show " secondary " optic atrophy. Their outlines are finely irregular and both are of a pale yellow colour. Otherwise the right fundus appears normal. The left fundus (Fig. 7) shows a circumscribed, slightly raised, pale area close to the superior temporal vessels. It is irregularly quadrilateral, appearing somewhat larger than the optic disc. Over it the usual fundus redness is replaced by a pallor, and some choroidal vessels can be seen in its depth. In places it is bordered by a little fine pigment and a clear rim that quickly fades into the surrounding fundus. It has the appearance of an almost flat tumour within the choroid, slightly raising the overlying retina. At the macula there are some fine irregular pigment deposits and fine exudates suggestive of circulatory disturbance. The phakos is probably a neurofibroma of the choroid. The bilateral secondary optic atrophy is probably due to similar tumours associated with the optic nerves. The patient cannot be X-rayed owing to his mental state.

Summary

Three cases of intra-ocular phakomata are recorded. The appearances of the retinal phakomata in the two patients with Bourneville's disease (tuberous sclerosis) are so characteristic that

diagnosis could be made if these were the only stigmata found. The diagnosis of the probable choroidal neurofibroma depends on the presence of very numerous, widely spread neurofibromata readily identified elsewhere, with bilateral secondary optic atrophy, epilepsy and amentia. On appearances alone its diagnosis could not be differentiated from other choroidal tumours.

Acknowledgments.—These investigations were conducted from the Institute of Ophthalmology, University of London. My thanks are due to the Dean (Mr. R. C. Davenport) and the Academic Board for the facilities provided. The patients were examined at Leavesden Hospital. I am most appreciative of the kindness and co-operation given me by the physician-superintendent, Dr. J. K. Watkin, and of the clinical help given me by Dr. W. M. McGrath. I have drawn freely on the clinical records and museum material of the hospital.

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ANNOTATIONS

Results and causes

To those who insist on putting first things first our title will be of the nature of an hypallage—the cart before the horse. In ophthalmology, especially in hospital practice, it is astonishing to what a variety of causes the O.P. is apt, in moments of expansion, to attribute the condition for which he or she seeks advice. Mothers often opine that a child's squint has resulted from attempts at copying some other squinter, either at home or among friends; and the same is also said of chorea. We recall an elderly male out-patient with a tarsal cyst who said it was hereditary. Perhaps it was, we did not feel called upon to question his statement.

There is really no knowing to what a patient will ascribe his condition. Tristram Shandy thought that the "asthma," from which he suffered, was due to skating against the wind in Flanders; but we do not believe that any author of a text-book on medicine has ever seriously included it among the causes of asthma. Sterne was, of course, a consumptive, and gives a very odd cause for a fit of laughter which brought on an attack of haemoptysis. It is not suitable for inclusion here, but will be found in the original (Tristram Shandy, Vol. VIII, Chapter 6, of the collected edition of Sterne's

works published in 1808). Lovers of Sir Walter, and there can be very few who are not, will remember the alarming consequence of the only occasion on which Dominie Sampson was ever known to laugh: his landlady miscarried!

Enough instances have been given. Those who think that the allusions we have drawn from fiction are too far fetched to be seriously considered, may yet one day in the course of their daily work, find something to make them alter their opinion.

Verba de praesenti

Our heading is defined as a law term meaning a promise made on the instant as a pledge for the future. The schoolmaster in effect makes use of it when he invites one of his young charges to come to his study to be caned. "I never thrashed a boy in a hackney coach before," said Mr. Squeers when he stopped to rest. "There's inconveniency in it, but the novelty gives it a sort of relish too."

One can hardly connect the castigation of Smike with ophthalmology; but any piece of work well done does give much satisfaction to the performer whatever his occupation may be. And the beginner, who for the first time, makes a really good section for the extraction of cataract may be pardoned if he experiences a satisfaction similar to that of Mr. Squeers, for a good piece of work he had not previously been able to accomplish. We fancy that most surgeons will know of instances where everything went wrong with the case in spite of perfection of handicraft. And all of us, including the victim, would rather put up with good visual results from an imperfect operation than bad results from the most perfect technique.

Even the most stoical patient is not exactly cheered by the prospect of an operation; and while it is natural and at the same time appropriate for the surgeon to do his best to allay depressive tendencies, we should be careful, if only in our own interests, to say nothing that will give the patient the impression that everything will go off well, as if all depended on the surgeon and nothing on the patient.

A distressing example of what we mean occurred many years ago when we were asked to assist at an operation for cataract. In this case one eye had already been operated on and the visual result was not very good; about 6/18 to the best of our recollection. As the patient was ascending the table he said in a gloomy tone, "I hope the result of this operation will be better than that of the last." The surgeon, to reassure him said, "Oh, it's going to be much better." At the time we thought that had we ourselves been called on to answer such a statement, we should not have said much more than "I hope so too." The operation went off perfectly satisfactorily but the eye did badly and had to be removed.

16th INTERNATIONAL CONGRESS OF OPHTHALMOLOGY

Tel.: Holborn 3474. *Office: 45, Lincoln's Inn Fields, London, W.C.2.*

Preliminary Notice

THEIR MAJESTIES THE KING AND QUEEN have been graciously pleased to grant their Patronage to the 16th International Congress of Ophthalmology, which, by decision of the International Council of Ophthalmology, will be held in London under the Presidency of Sir Stewart Duke-Elder, from July 17th—July 21st, 1950. This is the first International Congress to be held for thirteen years; it is hoped that it will be a great success and bring together a generation of ophthalmologists from many countries who have never yet met each other.

Any qualified medical practitioner may become a member of the Congress; others not so qualified who have rendered service to Ophthalmology and who are approved by the Council will be eligible to attend.

Relatives and friends of members will be welcomed as Associate Members. They will be eligible to attend the opening and closing ceremonies and all social functions.

The three official languages of the Congress are English, French and Spanish: any language, however, may be used.

The International Council have agreed upon two main discussions on the following subjects:

(*a*) The clinical and social aspects of heredity in ophthalmology.

(*b*) The role of the sympathetic system in the genesis of vascular hypertension and its effect upon the eye.

Those desirous of participating in the main discussions or reading papers on other ophthalmological subjects should communicate with Mr. Keith Lyle, International Congress of Ophthalmology, 45, Lincoln's Inn Fields, London, W.C.2.

Papers should not be longer than 2,500 words and will be read in résumé form, for which the author will be allowed 10 minutes. On their submission, each paper should be accompanied by a résumé of about 250 words translated (preferably) into the three official languages. Papers published before the Congress are not acceptable. As the number of papers submitted may be greater than can be accommodated, the Publications Committee reserves the right of rejection. In view of present printing difficulties, no guarantee can be given that any paper will be included in the Official Report of the Congress unless it is received 6 months before the date of the meeting (*i.e.*, by February, 1950).

In addition to papers, scientific demonstrations and exhibits will be arranged, and there will be scientific and industrial exhibitions. Contributions to all of these will be welcomed.

Further details, including information on the Social Programme, will be published from time to time.

FRANK W. LAW.
Secretary-General.

BOOK NOTICE

Strabismus. A Clinical Handbook. By GEORGE J. EPSTEIN. 254 pp., 76 figs. Philadelphia: The Blakiston Co. 1948. Price, 5 dollars.

A simple and comprehensive elementary book surveying the physiology and pathology of the extra-ocular muscles in their relation to strabismus. The anatomy and physiology of the muscles are first discussed and considerable emphasis laid on the physiology of binocular vision. The normal muscle balance is then described, and brief reference made to heterophoria. The main part of the volume deals with the clinical features of concomitant and paralytic strabismus. Non-operative methods of treatment, including orthoptic exercises are briefly noted, and the final section is devoted to a detailed discussion of the surgery of strabismus—the types of operation, their rationale, the appropriate surgical anatomy and the surgical techniques.

NOTES

International Congress of Ophthalmology

A LIST of doctors practising ophthalmology in Great Britain is being compiled for the new Indicia in connection with the forthcoming International Congress of Ophthalmology. Will all those who are *not* already members of the Faculty of Ophthalmologists or the Ophthalmological Society of the United Kingdom kindly submit their names, qualifications and addresses to the Honorary Secretary, Ophthalmological Society of the United Kingdom, 45, Lincoln's Inn Fields, London, W.C.2.

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Sociedad de Oftalmologia del Litoral

THE following officers of the Society during the next two years have been elected. *President*:—Dr. Juan Manuel Vila Ortiz; *Vice-President*:—Dr. Juan Maggi Zavalía; *Secretary*:—Dr. Máximo Carlos Soto; *Treasurer*:—Dr. Benito Bianco; *Vocales*:—Drs. Luis Woelflin, René Bazet, Mario Zurbriggen, Juan P. Allliani.